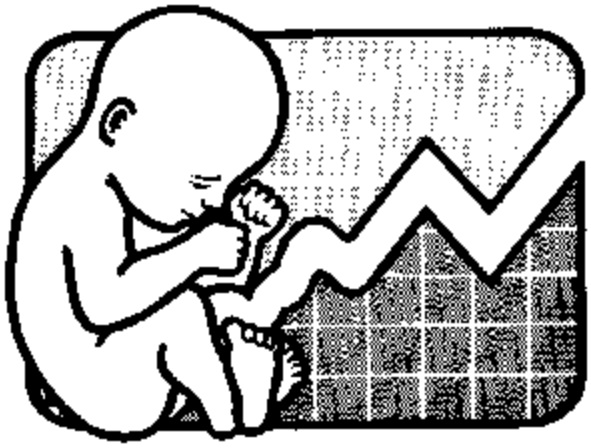
Arizona Department of Health Services

1991
Arizona Birth Defects Monitoring Program Report



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Jane Dee Hull
Governor
State of Arizona

James R. Allen, MD, MPH
Director
Arizona Department of Health Services

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1991 ARIZONA BIRTH DEFECTS MONITORING PROGRAM REPORT

Arizona Birth Defects Monitoring Program
Office of Chronic Disease Epidemiology
Bureau of Epidemiology and Disease Control Services
Arizona Department of Health Services

1400 West Washington Street, Ste. #127
Phoenix, Arizona 85007
(602) 542-7349

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by

Thomas E. Hughes, MA
Epidemiologist
Arizona Birth Defects Monitoring Program
Office of Chronic Disease Epidemiology

ACKNOWLEDGMENTS

Hoa Lien Tran .....Program Manager

Matilda Armenta .....Lead Abstractor

Laura Cook .....Abstractor

Elisa Duarte.....Secretary

Richard Porter.....Office Chief

Tim Flood, M.D. .....Medical Director

We thank the hospitals and health care providers of the state for their cooperation and active support of the Arizona Birth Defects Monitoring Program (ABDMP). Our special thanks go to the directors and employees of the Medical Record Departments of the hospitals and clinics, without whose help we could not accomplish our case finding and abstracting.

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EXECUTIVE SUMMARY

In 1991 there were 2,148 children with a reportable birth defect born to Arizona residents. During this period there were 68,040 live births and 409 still births in Arizona. This report presents 47 composite categories of birth defects developed by the Centers for Disease Control and Prevention (CDC). These categories represent the most serious defects, and they are the birth defects most frequently addressed in the scientific literature. Arizona's overall birth defect rate is 31.4 cases per 1,000 births. Pyloric stenosis, microcephalus, oral clefts, dislocation of hip, and Down syndrome were the most common birth defects.

Race/Ethnicity Patterns

Among Native Americans and Blacks microcephalus was the most common anomaly, while among Whites and Hispanics it was pyloric stenosis. Spina bifida was the most common neural tube defect (NTD) among all races; however, rates were highest among Hispanics. Down syndrome, a chromosomal defect, exhibited highest rates among Native Americans.

Age Patterns

Observed rates for all birth defects were highest among women 35 years of age and older. Down syndrome (Trisomy 21) rates increased with maternal age. Gastroschisis, an abdominal wall defect, showed highest rates among young mothers and decreased in incidence with maternal age.

County Patterns

For the first time, we present birth defects data by county. Cases were aggregated for the years 1986 through 1991 to provide numbers large enough for analysis. Gila county had the highest rate of congenital anomalies, whereas Greenlee and La Paz counties had the lowest rates. However, there was no statistically significant difference in overall rates between any of Arizona's 15 counties. Five sentinel defects (chromosomal defects, oral clefts, heart defects, abdominal wall defects, and neural tube defects) were examined by county.

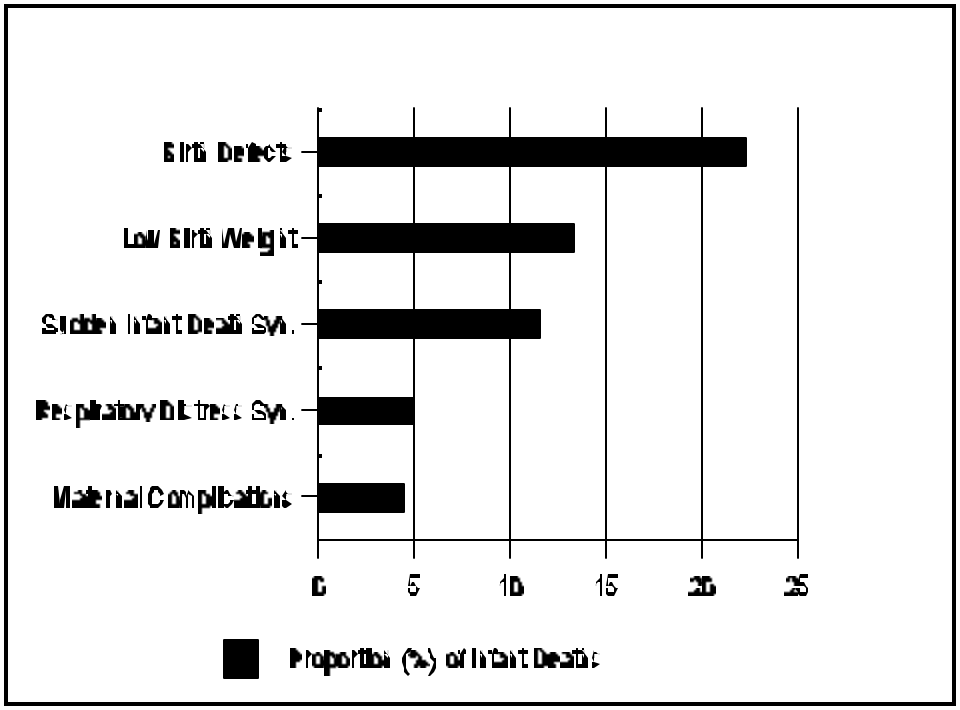
THE IMPORTANCE OF ARIZONA'S BIRTH DEFECTS REGISTRY

The need for a birth defects registry arises from multiple public health issues. The Arizona Birth Defects Monitoring Program (ABDMP) provides accurate counts used for prevention efforts, planning health services, and ongoing surveillance to monitor for trends and early detection of problems. Why is a registry necessary if birth defect information is collected on the birth certificate? Other systems for reporting birth defects, including birth certificates and hospital discharge data are often not accurate or complete. A recent study found that birth defects were reported on the birth certificate only 19% of the time. In addition, many defects were misclassified in the other reporting sources, while many babies were incorrectly identified as having a birth defect when in fact they did not.[1]

Economic Impact

The leading cause of infant mortality in the United States is birth defects (see Figure 1).[2] While it has been known that the cost of birth defects in the U.S. is enormous, past collection methods of anomalies have not provided accurate estimates of the economic cost. A recent study using California's population-based data (adjusted to provide national estimates) estimated the costs of the most clinically important structural birth defects in the United States. Estimates from this study found costs ranging from $75,000 to $503,000 per new case. The following are estimates for selected congenital

Figure 1. Leading Causes of Infant Mortality in the United States, 1995



anomalies: Down Syndrome ($451,000); the heart defect Truncus Arteriosus, ($503,000) and Spina Bifida ($294,000).[3]

Human Cost

Each year approximately 2,400 children are born with a birth defect in Arizona. An estimated 150,000 babies are born with serious birth defects in the United States every year.[4] While we know the economic costs associated with birth defects are high, there is no way to measure the human and societal cost.

While some defects such as fetal alcohol syndrome and German measles are preventable, many defects are caused by unknown teratogens. The search for causes of birth defects is a difficult process. If Arizona is to ensure its children a healthy future, we must continue to search for the causes of congenital anomalies, and birth defect registries are a vital first step in reducing birth defects. The documentation of baseline birth defect rates in Arizona provides the starting point against which we can measure successful interventions.

DATA SOURCES AND PROCEDURES

The ABDMP is a statewide, population-based, active surveillance program, pursuant to ARS §36-133 which mandates the surveillance of chronic diseases, including birth defects. Trained ABDMP staff collect data from 63 reporting sources: 58 hospitals, including Phoenix Children's Hospital; 1 center providing genetics services; 4 clinics of the state Children's Rehabilitative Services; and the state Office of Vital Records. Ascertainment procedures used by the ABDMP are nearly identical to those used by the California Birth Defects Monitoring Program and the US Centers for Disease Control's Metropolitan Atlanta Congenital Defects Program (MACDP).

Sources of data at hospitals include the disease index; labor and delivery log; nursery log; newborn intensive care log; pediatric log; pathology/autopsy log. Not all sources are available at each hospital. The medical record numbers of potential cases are identified through a review of the hospital's disease index and various logs. This process is called case finding. Next, the medical records of possible cases are pulled and reviewed to determine which of the potential cases actually meet the case definition. An abstract of the medical record then is completed for each reportable case. In order to find the birth defect cases born in 1991, ABDMP staff reviewed more than 10,000 medical records, identified reportable cases, and excluded those not meeting the case definition.

In addition to the hospital sources, Certificates of Birth, Death, and Fetal Death that indicate a birth defect are reviewed and matched against cases listed in the registry. Medical records then are requested from the reporting hospital on those children not previously identified from other sources and if the condition(s) reported meet the case definition, pertinent information is abstracted for the registry. If the nature of a defect diagnosed in the first year of life is more precisely diagnosed later in the child's life and this information is contained in the chart at the time of our review (which occurs 2-4 years after the fact) then the more precise diagnosis is used.

The abstracts of cases identified from multiple sources are compared, merged, and added to the registry. Inconsistencies, differences and/or conflicting data are resolved before being entered into the ABDMP system.

ABDMP staff assign a six-digit classification code to each defect. The classification system is CDC's modification of the British Pediatric Association (BPA) Classification of Disease. This coding system is similar to the International Classification of Disease (ICD). The staff collect diagnostic information on birth defects that fall primarily within the range of ICD-9-CM Codes 740.00-759.99. The system of codes is hierarchical: the more digits in the code, the more precise the diagnosis. ABDMP staff always

code the data at the most precise level possible.

CASE DEFINITION

The following are the criteria for inclusion in the Birth Defects Monitoring Program case file:

A. The mother's place of residence at the time of birth must be in Arizona.

B. The child must have a structural, genetic, or biochemical birth defect, or other specified birth outcome that can adversely affect an infant's health and development (most, but not all, are listed in ICD-9-CM 740.0-759.9).

C. The defect must be diagnosed, or signs and symptoms of a potential defect recognized, within the first year of life.

D. Stillborn infants are included if they have a reportable birth defect.

E. The date of birth (or delivery for stillbirths>19 weeks) is on or after January 1, 1986.

INTERPRETING THE DATA

The tables and figures presented in this report represent data collected on birth defects in Arizona for the period 1986 to 1991. Each table presents reported counts, rates and confidence intervals on selected congenital anomalies. Below is an explanation of how counts, rates, and confidence intervals were calculated.

Counts

The counts, sometimes called cases, represent the number of children who were diagnosed with a particular reportable birth defect within the first year of life. Children born with more than one reportable defect, as often occurs, may be part of the counts across multiple rows.

Rates

Incidence rates of birth defects were calculated by dividing the number of children with a particular reportable defect by the total number of live births (and in some cases live births plus fetal deaths) for the specific year of interest and then multiplying by 10,000. For example, there were 84 cases of Down Syndrome in 1991 and 68,040 live births in 1991, the rate is calculated as such: 84/68,040*10,000 = 12.34 cases of Down Syndrome per 10,000 live births.

Confidence Intervals

The confidence intervals shown in the tables and figures are provided to give information about the estimate of the rate. Confidence intervals presented in this report are the 99 percent Poisson confidence intervals. The confidence intervals tell one that the true rate should be contained in this interval 99 percent of the time. For example, Down Syndrome occurs at a rate of 12.34 per 10,000 live births. The lower and upper bounds of the point estimate in this case are 9.1 and 16.3, respectively. Thus, one can say with 99 percent certainty that the true rate of Down Syndrome is between 9.1 and 16.3 cases per 10,000 live births.

Small Numbers and a Note Of Caution

While the intent of these data is to provide the reader with useful information on birth defects in Arizona, an equally important point is not to mislead data users. Therefore, it is important to stress that rates, confidence intervals, or any other analysis based on fewer than 10 reported cases cannot be considered statistically reliable.

STATE PROFILE

State Data

This is the sixth annual report of data compiled by the ABDMP in its mission to collect and analyze information on children with birth defects and to provide data for the study of causes of birth defects in Arizona.

Tables and Figures

Table 1 presents data on 47 selected congenital anomalies by race for 1991. Table 2 looks at all reportable birth defects for both live births and fetal deaths. Fetal deaths include therapeutic abortions and still born babies with a reportable congenital defect if the estimated gestational age is greater than 19 weeks. Table 3 displays birth defect rates by year for 1986 through 1991. A new addition to this report is figure 2 that looks at trends for selected congenital anomalies.

County and Race/Ethnicity

An expanded look at birth defects and race/ethnicity follows the state profile. Likewise, analysis of county level data is presented later in this report under the heading County Profiles. As always, the Arizona Birth Defects Monitoring Program welcomes comments, questions, and observations.

Table 1
Arizona Birth Defects Monitoring Program
Congenital Anomalies - Arizona 1991
Incidence Rates per 10,000 Live Births and Fetal Deaths

CODE	DEFECT GROUP	TOTAL	RATE	WHITE	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
A00	CENTRAL NERVOUS SYSTEM												
A01	Anencephaly	17	2.48	10	2.60	2	.98	0	0.00	3	4.92	2	19.78
A02	Spina Bifida w/ Hydrocephaly	21	3.06	10	2.60	6	2.94	2	7.81	3	4.92	0	0.00
A03	Spina Bifida w/o Hydrocephaly	13	1.89	03	.78	6	2.94	0	0.00	2	3.28	2	19.78
A13	Encephalocele	14	2.04	10	2.60	3	1.47	0	0.00	1	1.64	0	0.00
A15	Hydrocephaly	46	6.72	28	7.28	8	3.92	1	3.90	9	14.76	0	0.00
A16	Microcephalus	120	17.53	43	11.19	30	14.73	11	43.00	35	57.42	1	9.89
B00	EYE AND EAR												
B03	Glaucoma	2	0.29	1	0.26	1	0.49	0	0.00	0	0.00	0	0.00
B04	Cataract	10	1.46	7	1.82	3	1.47	0	0.00	0	0.00	0	0.00
B51	Anophthalmia	5	0.73	5	1.30	0	0.00	0	0.00	0	0.00	0	0.00
B52	Microphthalmia	29	4.23	14	3.64	10	4.91	0	0.00	5	8.20	0	0.00
B54	Ear Anomaly w/ hearing loss	65	9.49	26	6.76	26	12.76	0	0.00	12	19.68	1	9.89
D00	CARDIAC												
D01	Truncus Arteriosus	6	0.87	2	0.52	3	1.47	0	0.00	0	0.00	1	9.89
D02	Transposition of great vessels	26	3.79	13	3.38	11	5.40	2	7.81	0	0.00	0	0.00
D03	Tetralogy of Fallot	22	3.21	14	3.64	5	2.45	2	7.81	1	1.64	0	0.00
D04	Single ventricle	1	0.14	0	0.00	1	0.49	0	0.00	0	0.00	0	0.00
D51	Aortic stenosis	17	2.48	10	2.60	6	2.94	1	3.90	0	1.60	0	0.00
D52	Hypoplastic left heart	11	1.60	8	2.08	3	1.47	0	0.00	0	1.60	0	0.00
D53	Tot. anomal. pulm. ven. return	11	1.60	7	1.82	2	0.98	0	0.00	2	3.28	0	0.00
E00	RESPIRATORY												
E01	Choanal atresia	5	0.73	1	0.26	2	0.98	0	0.00	1	1.64	1	9.89
E06	Agenesis of lung	50	7.30	27	7.02	13	6.38	3	11.72	5	8.20	2	19.78
F00	OROFACIAL AND GASTROINTESTINAL												
F01	Cleft palate	31	4.52	19	4.94	9	4.42	0	0.00	3	4.92	0	0.00
F01	Cleft lip w&wo cleft palate	80	11.68	41	10.67	24	11.78	1	3.90	13	21.32	1	9.89
F03	Pyloric stenosis	148	21.62	80	20.82	52	25.53	4	15.63	12	19.68	0	0.00
F09	Tracheo-esophageal fistula	15	2.19	12	3.12	2	0.98	1	3.90	0	0.00	0	0.00

Incidence rates based on counts of less than 10 events are not statistically reliable.

Table 1 Continued
Arizona Birth Defects Monitoring Program
Congenital Anomalies - Arizona 1991
Incidence Rates per 10,000 Live Births and Fetal Deaths

CODE	DEFECT GROUP	TOTAL	RATE	WHITE	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
F00	OROFACIAL AND GASTROINTESTINAL												
F14	Stenosis/atresia of duodenum	6	0.87	4	1.04	1	0.49	0	0.00	1	1.64	0	0.00
F15	Stenosis/atresia of sm. intest	9	1.31	3	0.78	4	1.96	1	3.90	1	1.64	0	0.00
F16	Stenosis/atresia of rectum	38	5.55	26	6.76	7	3.43	1	3.90	2	3.28	2	19.78
F17	Hirschsprung's disease	13	1.89	5	1.30	6	2.94	1	3.90	1	1.64	0	0.00
F18	Malrotation of intestine	14	2.04	11	2.86	1	0.49	0	0.00	2	3.28	0	0.00
F21	Biliary atresia	6	0.87	3	0.78	0	0.00	1	3.90	1	1.64	1	9.89
H00	GENITO-URINARY												
H01	Renal agenesis	37	5.40	22	5.72	9	4.42	1	3.90	3	4.92	2	19.78
H06	Obstruction of kidney/ureter	103	15.04	54	14.05	37	18.17	4	15.63	6	9.84	2	19.78
H09	Bladder or urethra obstruction	8	1.16	4	1.04	3	1.47	1	3.90	0	0.00	0	0.00
J00	MUSCULOSKELETAL												
J02	Curvature of spine	35	5.11	18	4.68	12	5.89	1	3.90	4	6.56	0	0.00
J03	Dislocation of hip	103	15.04	58	15.09	31	15.22	1	3.90	11	18.04	2	19.78
J11	Arthrogryposis multiplex cong.	12	1.75	8	2.08	3	1.47	0	0.00	1	1.64	0	0.00
J51	Complete absence upp/low limb	2	0.29	2	0.52	0	0.00	0	0.00	0	0.00	0	0.00
J52	Phocomelia of Limb	1	0.14	1	0.26	0	0.00	0	0.00	0	0.00	0	0.00
K05	Amniotic Bands	10	1.46	4	1.04	5	2.45	0	0.00	1	1.64	0	0.00
N01	Diaphragmatic hernia	23	3.36	11	2.86	7	3.43	2	7.81	3	4.92	0	0.00
N02	Omphalocele	21	3.06	10	2.60	4	1.96	3	11.72	3	4.92	1	9.89
N04	Gastroschisis	36	5.25	20	5.20	14	6.87	1	3.90	1	1.64	0	0.00
R00	SYNDROMES												
R01	Down Syndrome (Trisomy 21)	84	12.27	38	9.89	30	14.73	2	7.81	12	19.68	2	19.78
R02	Patau Syndrome (Trisomy 13)	6	0.87	3	0.78	2	0.98	0	0.00	0	0.00	1	9.89
R03	Edwards Syndrome (Trisomy 18)	13	1.89	6	1.56	5	2.45	0	0.00	1	1.64	1	9.89
S02	Fetal Alcohol Syndrome	27	3.94	2	0.52	1	0.49	3	11.72	21	34.45	0	0.00
W03	Conjoined twins	2	0.29	2	0.52	0	0.00	0	0.00	0	0.00	0	0.00

Incidence rates based on counts of less than 10 events are not statistically reliable.

Table 2
Arizona Birth Defects Monitoring Program [1,2]
Birth Defects by County of Residence, 1991

STATE, COUNTY	LIVE BIRTHS W/DEFECTS		STILL BIRTHS W/ DEFECTS		LIVE AND STILL W/ DEFECTS		NUMBER OF DEFECTS OF LIVE BIRTHS		NUMBER OF DEFECTS OF STILL BIRTHS	
	Number	% OF LB	Number	% OF SB	Number	% TOT.	Number	AVG Number	Number	AVG Number
ARIZONA	2082	3.06	66	16.14	2148	3.14	7914	3.80	292	4.42
APACHE COUNTY	59	3.40	2	28.75	61	3.50	296	5.02	5	2.50
COCHISE COUNTY	41	2.53	2	28.57	43	2.64	125	3.05	27	13.50
COCONINO COUNTY	54	2.77	1	10	55	2.80	154	2.85	4	4.00
GILA COUNTY	21	3.28	0	0	21	3.25	162	7.71	0	0
GRAHAM COUNTY	16	3.62	0	0	16	3.59	84	5.25	0	0
GREENLEE COUNTY	3	1.95	0	0	3	1.94	6	2	0	0
LA PAZ COUNTY	5	2.72	0	0	5	2.70	42	8.40	0	0
MARICOPA COUNTY	1237	3.10	34	14.41	1271	3.17	4393	3.55	152	4.47
MOHAVE COUNTY	34	2.18	1	25.00	35	2.24	112	3.29	1	1
NAVAJO COUNTY	58	3.10	2	16.67	60	3.18	262	4.52	2	1
PIMA COUNTY	353	3.12	19	27.14	372	3.27	1518	4.30	79	4.16
PINAL COUNTY	59	2.79	0	0	59	2.76	278	4.71	0	0
SANTA CRUZ COUNTY	26	3.35	0	0	26	3.34	100	3.85	0	0
YAVAPAI COUNTY	34	2.77	4	30.77	38	3.06	139	4.09	21	5.25
YUMA COUNTY	82	3.22	1	5.88	83	3.24	243	2.96	1	1.00

[1]Total number of live births in Arizona for 1991 = 68,040
[2]Total number of fetal deaths in Arizona for 1991 = 409

Table 3
Arizona Birth Defects Monitoring Program
Incidence Rates Per 1,000 Live Births and Fetal Deaths[1]
Arizona, 1991

CODE/CONDITION (1)							
		1986	1987	1988	1989	1990	1991
A01 Anencephaly	Cases Rate CI	22 0.35 0.19-0.60	17 0.26 0.12-0.48	18 0.27 0.13-0.48	18 0.27 0.13-0.48	16 0.23 0.11-0.43	17 0.25 0.12-0.45
A02 S.B. with Hydrocephaly	Cases Rate CI	26 0.42 0.24-0.69	24 0.37 0.20-0.62	19 0.28 0.14-0.50	22 0.33 0.17-0.55	23 0.33 0.18-0.56	21 0.31 0.16-0.53
A03 S.B. without Hydrocephaly	Cases Rate CI	10 0.16 0.06-0.35	11 0.17 0.06-0.35	6 0.09 0.02-0.23	16 0.24 0.11-0.44	16 0.23 0.11-0.43	13 0.19 0.08-0.37
A13 Encephalocele	Cases Rate CI	10 0.16 0.06-0.35	8 0.12 0.03-0.29	14 0.21 0.09-0.40	5 0.07 0.02-0.21	13 0.19 0.08-0.37	14 0.20 0.09-0.39
A15 Hydrocephaly	Cases Rate CI	34 0.55 0.34-0.85	41 0.64 0.41-0.95	48 0.72 0.48-1.04	44 0.65 0.43-0.95	52 0.75 0.51-1.06	46 0.67 0.44-0.97
A16 Microcephalus	Cases Rate CI	30 0.49 0.29-0.77	60 0.94 0.65-1.30	70 1.06 0.76-1.43	109 1.61 1.17-1.96	118 1.70 1.33-2.15	120 1.75 1.37-2.21
B03 Glaucoma	Cases Rate CI	2 0.03 0.04-0.15	7 0.10 0.03-0.26	4 0.06 0.00-0.19	5 0.07 0.02-0.21	4 0.06 0.01-0.18	2 0.03 0.00-0.14

(1) See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.

Table 3 Continued
Arizona Birth Defects Monitoring Program
Incidence Rates Per 1,000 Live Births and Fetal Deaths[1]
Arizona, 1991

CODE/CONDITION (1)							
		1986	1987	1988	1989	1990	1991
B04 Cataract	Cases Rate CI	8 0.13 0.04-0.30	7 0.10 0.03-0.26	7 0.10 0.03-0.26	15 0.22 0.10-0.42	24 .35 0.19-0.57	10 0.15 0.05-0.31
B51 Anophthalmia	Cases Rate CI	6 0.09 0.02-0.25	1 0.01 0.00-0.11	3 0.04 0.00-0.16	5 0.07 0.02-0.21	7 0.10 0.03-0.25	5 0.07 0.02-0.21
B52 Microphthalmia	Cases Rate CI	10 0.16 0.06-0.35	24 0.37 0.20-0.62	21 0.31 0.16-0.54	19 0.28 0.14-0.50	24 0.35 0.19-0.57	29 0.42 0.25-0.67
B54 Hearing loss w/ear anomaly	Cases Rate CI	33 0.53 0.32-0.83	59 0.92 0.64-1.28	34 0.51 0.31-0.79	50 0.74 0.50-1.06	59 0.85 0.59-1.18	65 0.95 0.67-1.30
D01 Truncus Arteriosus	Cases Rate CI	4 0.06 0.01-0.20	10 0.15 0.05-0.33	9 0.13 0.04-0.30	9 0.13 0.05-0.30	6 0.09 0.02-0.23	6 0.09 0.02-0.23
D02 Transposition of Great Vessels	Cases Rate CI	32 0.52 0.31-0.81	26 0.40 0.23-0.66	26 .39 0.22-0.64	33 0.49 0.30-0.75	28 0.40 0.23-0.65	26 0.38 0.21-0.62
D03 Tetralogy of Fallot	Cases Rate CI	15 0.24 0.11-0.46	18 0.28 0.13-0.50	29 0.43 0.25-0.69	23 0.34 0.19-0.57	27 0.39 0.22-0.63	22 0.32 0.17-0.54

(1) See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.

Table 3 Continued
Arizona Birth Defects Monitoring Program
Incidence Rates Per 1,000 Live Births and Fetal Deaths[1]
Arizona, 1991

CODE/CONDITION (1)							
		1986	1987	1988	1989	1990	1991
D04 Single Ventricle	Cases Rate CI	2 0.03 0.00-0.15	4 0.06 0.01-0.19	5 0.07 0.01-0.21	4 0.06 0.01-0.19	6 0.09 0.02-0.23	1 0.01 0.00-0.11
D51 Aortic Stenosis	Cases Rate CI	8 0.13 0.04-0.30	15 0.23 0.10-0.44	17 0.25 0.12-0.46	25 0.37 0.21-0.61	17 0.25 0.12-0.45	17 0.25 0.12-0.45
D52 Hypoplastic Left Heart	Cases Rate CI	9 0.14 0.05-0.32	16 0.25 0.11-0.46	8 0.12 0.03-0.28	16 0.24 0.11-0.44	19 0.28 0.14-0.48	11 0.16 0.06-0.33
D53 Total Anomalous Pulmonary Venous Return	Cases Rate CI	5 0.08 0.17-0.23	5 0.07 0.01-0.22	13 0.19 0.08-0.38	17 0.25 0.12-0.46	13 0.19 0.08-0.37	11 0.16 0.06-0.33
E01 Choanal Atresia	Cases Rate CI	6 0.09 0.24-0.25	10 0.15 0.05-0.33	10 0.15 0.05-0.32	16 0.24 0.11-0.44	6 0.09 0.02-0.23	5 .07 0.02-0.21
E06 Agenesis of Lung	Cases Rate CI	25 0.40 0.22-0.67	44 0.69 0.45-1.00	32 0.48 0.29-0.75	42 0.62 0.40-0.92	49 0.71 0.47-1.01	50 0.73 0.49-1.04
F01 Cleft Palate	Cases Rate CI	39 0.63 0.40-0.95	46 0.72 0.47-1.04	36 0.54 0.33-0.82	43 0.64 0.41-0.93	38 0.55 0.35-0.82	31 0.45 0.27-0.71

(1) See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.

Table 3 Continued
Arizona Birth Defects Monitoring Program
Incidence Rates Per 1,000 Live Births and Fetal Deaths[1]
Arizona, 1991

CODE/CONDITION (1)							
		1986	1987	1988	1989	1990	1991
F02 Cleft Lip with and without Cleft Palate	Cases Rate CI	77 1.25 0.91-1.67	80 1.25 0.92-1.66	91 1.37 1.03-1.79	90 1.33 1.00-1.74	97 1.40 1.06-1.81	80 1.17 0.86-1.55
F08 Pyloric Stenosis	Cases Rate CI	108 1.76 1.35-2.25	135 2.11 1.67-2.63	134 2.03 1.60-2.52	122 1.81 1.41-2.27	116 1.68 1.30-2.12	148 2.16 1.73-2.66
F09 TE Fistula, or Esophageal Atresia, or both	Cases Rate CI	19 0.31 0.15-0.54	16 0.25 0.11-0.46	19 0.28 0.14-0.50	18 0.27 0.13-0.48	19 0.27 0.14-0.48	15 0.22 0.10-0.41
F14 Stenosis/Atresia of Duodenum	Cases Rate CI	5 0.08 0.01-0.23	15 0.07 0.01-0.22	11 0.16 0.06-0.34	10 0.15 0.05-0.32	10 0.14 0.05-0.31	6 0.09 0.02-0.23
F15 Stenosis/Atresia of Small Intestine	Cases Rate CI	18 0.29 0.14-0.52	12 0.18 0.07-0.37	13 0.19 0.08-0.38	16 0.24 0.11-0.44	16 0.23 0.11-0.43	9 0.13 0.05-0.29
F16 Stenosis/Atresia of Rectum or Anus	Cases Rate CI	27 0.44 0.25-0.71	26 0.40 0.23-0.66	27 0.40 0.23-0.66	35 0.52 0.32-0.79	35 0.51 0.31-0.78	38 0.56 0.35-0.83
F17 Hirschsprung's Disease	Cases Rate CI	11 0.17 0.07-0.37	10 0.15 0.05-0.33	10 0.15 0.05-0.32	7 0.03 0.03-0.25	13 0.19 0.08-0.37	13 0.19 0.08-0.37

(1) See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.

Table 3 Continued
Arizona Birth Defects Monitoring Program
Incidence Rates Per 1,000 Live Births and Fetal Deaths[1]
Arizona, 1991

CODE/CONDITION (1)							
		1986	1987	1988	1989	1990	1991
F18 Malrotation of Intestine	Cases Rate CI	10 0.16 0.06-0.35	10 0.15 0.05-0.33	16 0.24 0.11-0.44	14 0.21 0.09-0.40	16 0.23 0.11-0.43	14 0.20 0.09-0.39
F21 Biliary Atresia	Cases Rate CI	2 0.03 0.00-0.15	1 0.01 0.00-0.11	3 0.04 0.00-0.16	5 0.07 0.02-0.21	4 0.06 0.01-0.18	6 0.09 0.02-0.23
H01 Renal Agenesis	Cases Rate CI	21 0.34 0.18	27 0.42 0.24-0.68	23 0.34 0.18-0.58	43 0.64 0.41-0.93	33 0.48 0.29-0.74	37 0.54 0.34-0.82
H06 Obstruction Kidney/Ureter	Cases Rate CI	37 0.60 0.37-0.91	71 1.11 0.80-1.50	64 0.97 0.68-1.32	90 1.33 1.00-1.74	94 1.36 1.02-1.76	103 1.50 1.15-1.93
H09 Bladder or Urethra Obstruction	Cases Rate CI	8 0.13 0.04-0.30	12 0.18 0.07-0.37	9 0.13 0.04-0.30	7 0.10 0.03-0.25	3 0.04 0.00-0.16	8 0.12 0.04-0.27
J02 Scoliosis/Lordosis	Cases Rate CI	15 0.24 0.11-0.46	13 0.20 0.08-0.40	19 0.28 0.14-0.50	35 0.52 0.32-0.79	48 0.69 0.46-1.00	35 0.51 0.32-0.78
J03 Dislocation of Hip	Cases Rate CI	87 1.42 1.05-1.86	101 1.58 1.20-2.03	68 1.03 1.20-2.03	91 1.35 1.01-1.76	105 1.52 1.16-1.76	103 1.50 1.15-1.93

(1) See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.

Table 3 Continued
Arizona Birth Defects Monitoring Program
Incidence Rates Per 1,000 Live Births and Fetal Deaths[1]
Arizona, 1991

CODE/CONDITION (1)							
		1986	1987	1988	1989	1990	1991
J11 Arthrogryposis Multiplex Congenital	Cases Rate CI	9 0.14 0.05-0.32	22 0.34 0.18-0.58	13 0.19 0.08-0.38	33 0.49 0.30-0.75	34 0.49 0.30-0.75	12 0.18 0.07-0.35
J51 Complete absence of upper or lower limb	Cases Rate CI	2 0.03 0.00-0.15	0 0.00 0.00-0.00	1 0.01 0.00-0.11	3 0.04 0.00-0.16	3 .04 0.00-0.16	2 0.03 0.00-0.14
J52 Phocomelia of limb	Cases Rate CI	3 0.04 0.00-0.18	2 0.03 0.00-0.14	2 .03 0.00-0.14	1 0.01 0.00-0.25	1 0.01 0.00-0.11	1 0.01 0.00-0.11
K05 Amniotic Bands	Cases Rate CI	4 0.06 0.01-0.20	4 .06 0.01-0.19	9 0.14 0.05-0.32	8 0.11 0.04-0.28	14 0.20 0.09-0.39	10 0.15 0.05-0.31
N01 Diaphragmatic	Cases Rate CI	13 0.21 0.09-0.41	18 0.28 0.13-0.50	20 0.30 0.15-0.52	23 0.34 0.19-0.57	28 0.40 0.23-0.65	23 0.34 0.18-0.56
N02 Omphalocele	Cases Rate CI	10 0.16 0.06-0.35	14 0.21 0.09-0.42	17 0.25 0.12-0.46	10 0.15 0.05-0.32	21 0.30 0.16-0.52	21 0.31 0.16-0.53
N04 Gastroschisis	Cases Rate CI	19 0.31 0.15-0.54	18 0.28 0.13-0.50	19 0.28 0.14-0.50	19 0.28 0.14-0.50	21 0.30 0.16-0.52	36 0.53 0.33-0.80

(1) See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.

Table 3 Continued
Arizona Birth Defects Monitoring Program
Incidence Rates Per 1,000 Live Births and Fetal Deaths[1]
Arizona, 1991

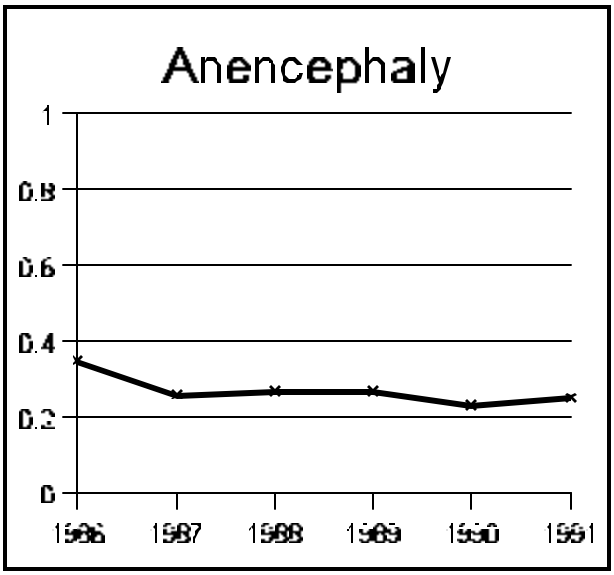
CODE/CONDITION (1)							
		1986	1987	1988	1989	1990	1991
R01 Down Syndrome (Trisomy 21)	Cases Rate CI	64 1.04 0.73-1.43	61 0.95 0.67-1.32	74 1.12 0.81-1.50	66 0.98 0.70-1.33	73 1.05 0.76-1.42	84 1.23 0.91-1.62
R02 Patau Syndrome (Trisomy 13)	Cases Rate CI	9 0.14 0.05-0.32	4 0.06 0.01-0.19	3 0.04 0.00-0.16	4 0.06 0.01-0.19	11 0.16 0.06-0.33	6 0.09 0.02-0.23
R03 Edwards Syndrome (Trisomy 18)	Cases Rate CI	11 0.17 0.07-0.37	17 0.26 0.12-0.48	13 0.19 0.08-0.38	10 0.15 0.05-0.32	15 0.22 0.10-0.41	13 0.19 0.08-0.37
S02 Fetal Alcohol Syndrome	Cases Rate CI	9 0.14 0.05-0.32	25 0.39 0.21-0.64	12 0.18 0.07-0.36	21 0.31 0.16-0.53	22 0.32 0.17-0.54	27 0.39 0.23-0.64
W03 Conjoined Twins	Cases Rate CI	2 0.03 0.00-0.15	0 0.00 0.00-0.00	2 0.03 0.00-0.14	0 0.00 0.00-0.00	2 0.03 0.00-0.13	2 0.03 0.00-0.14

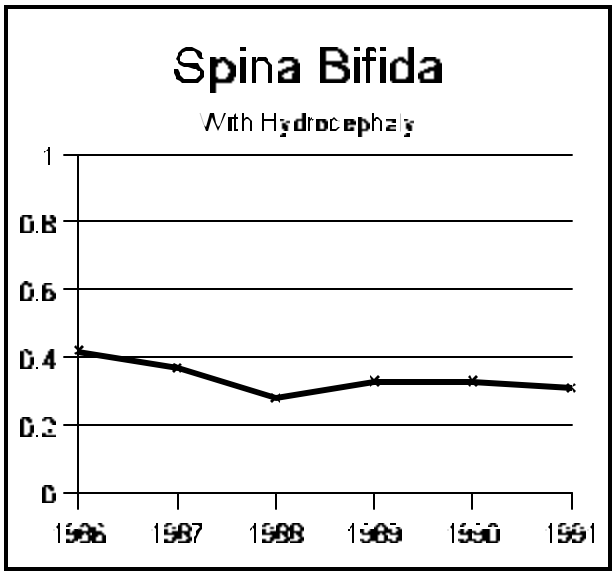
(1) See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.
Cases = Number of live births and fetal deaths >= 20 weeks.

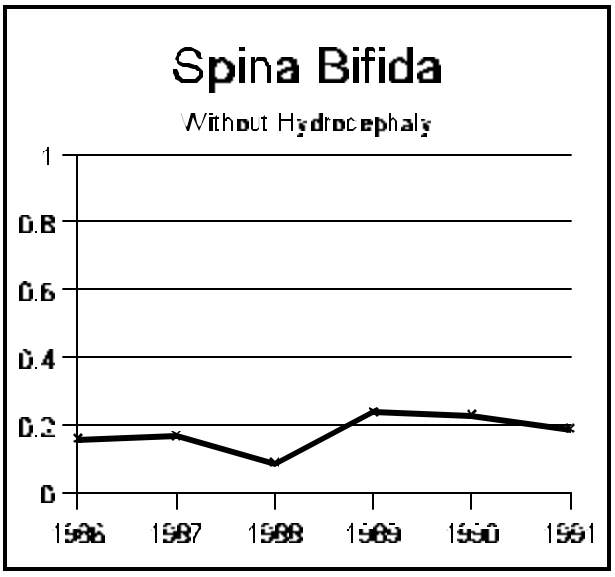
[1]Denominators -

1986= 61,203; 1987= 63,742; 1988= 65,981; 1989= 67,498; 1990= 69,245; 1991= 68,449

Figure 2. Trends of Selected Congenital Anomalies: Incident Rates per 1,000 Live Births & Fetal Deaths, Arizona







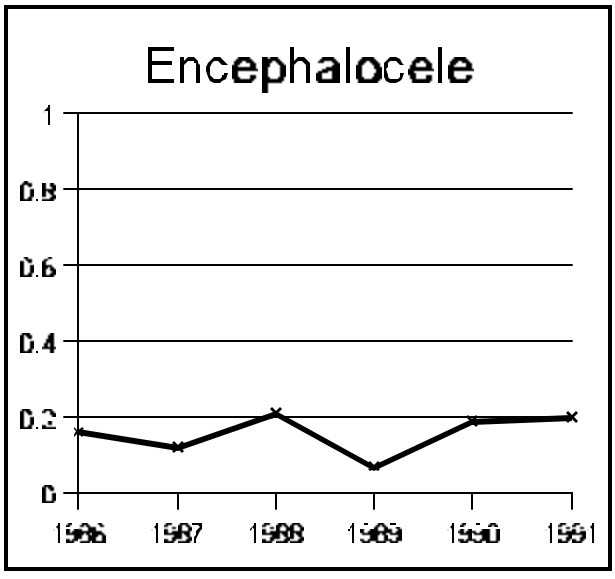
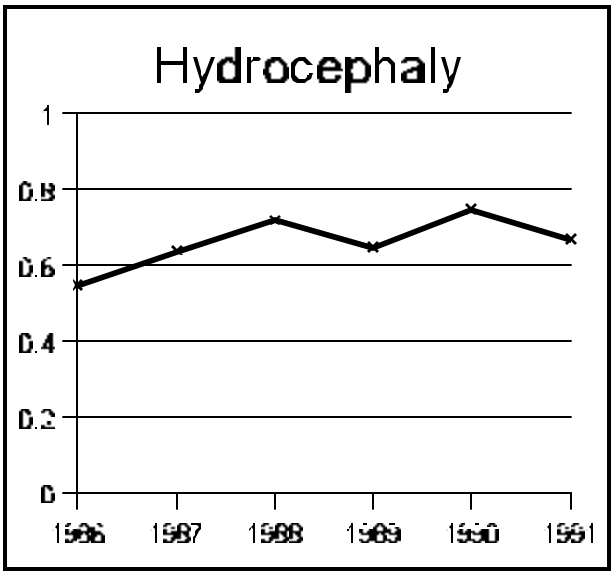
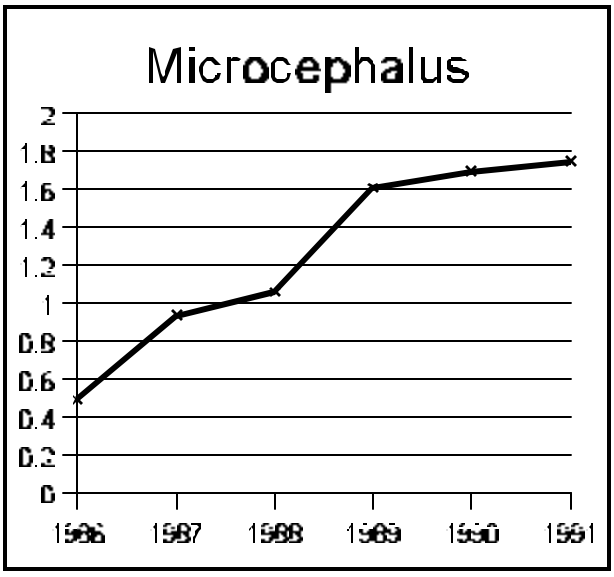
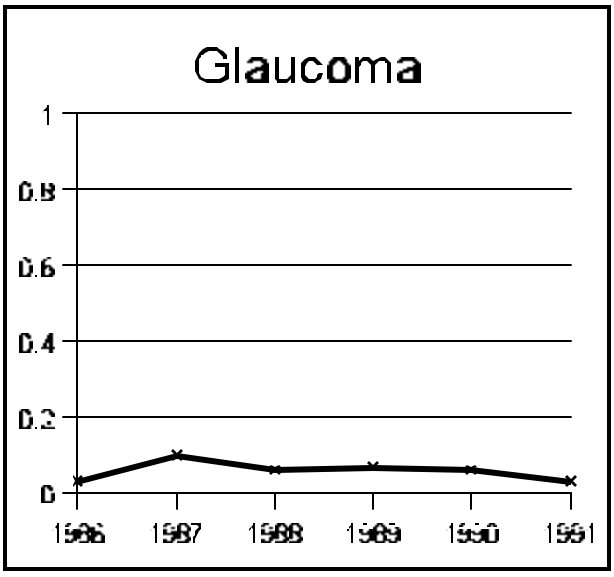


Figure 2. Trends of Selected Congenital Anomalies: Incident Rates
per 1,000 Live Births & Fetal Deaths, Arizona







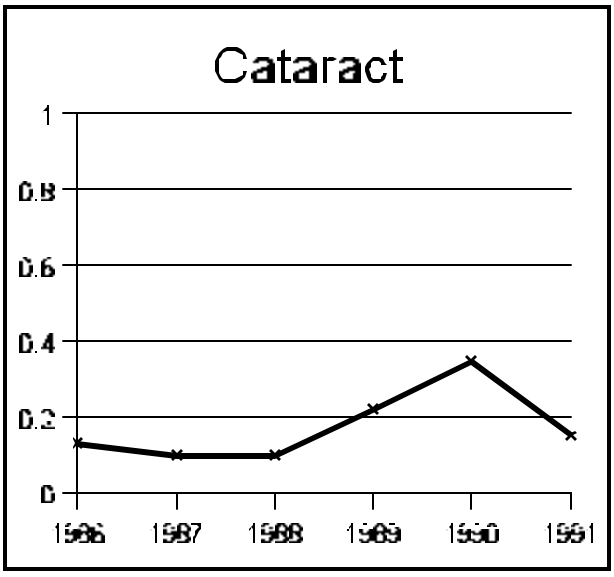
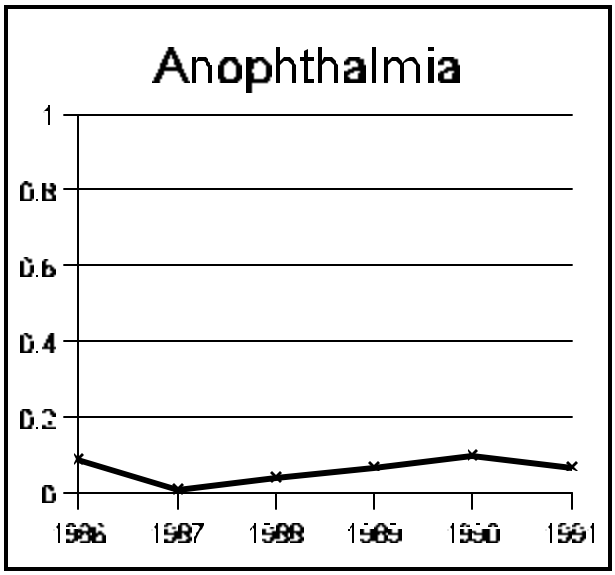
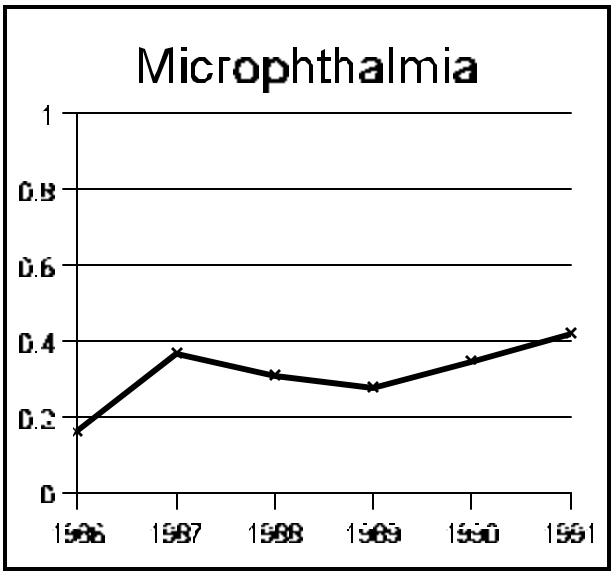
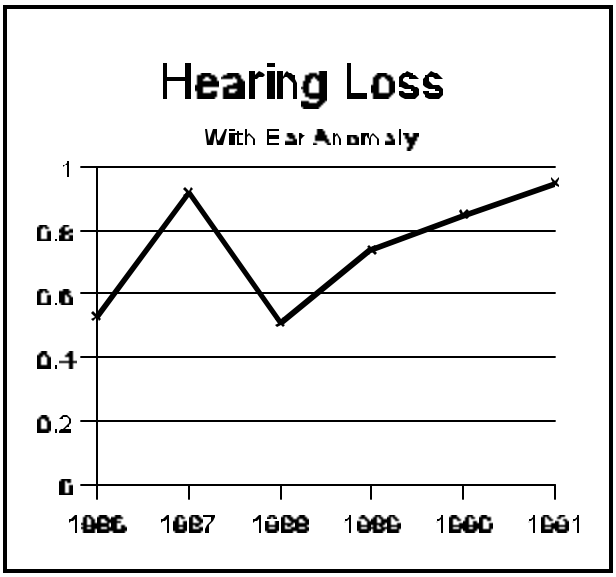


Figure 2. Trends of Selected Congenital Anomalies: Incident Rates per 1,000 Live Births & Fetal Deaths, Arizona







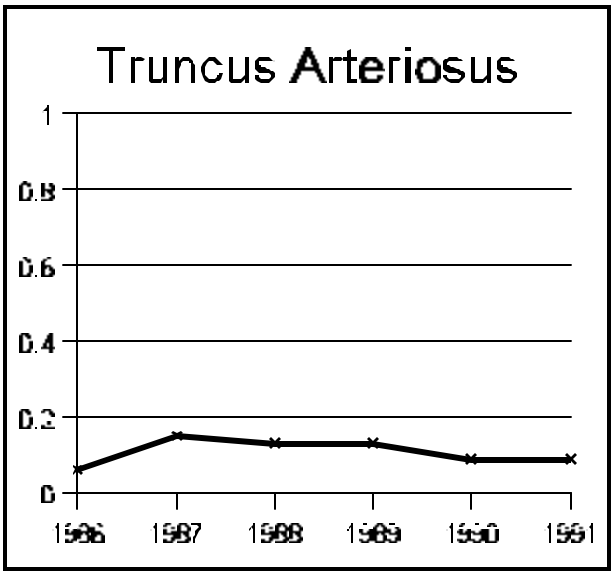
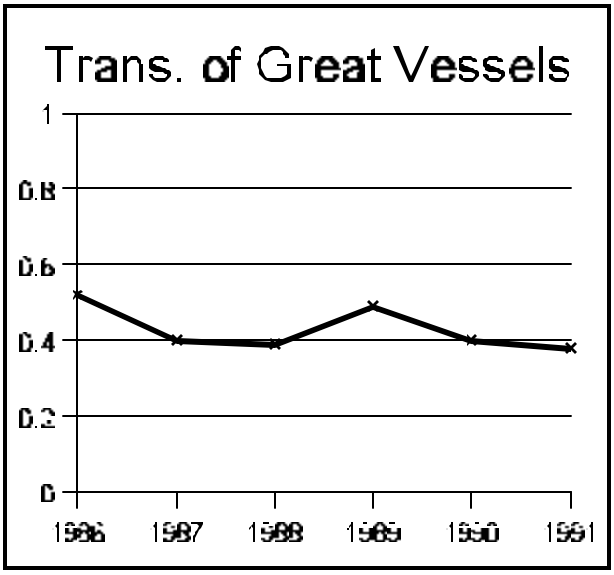
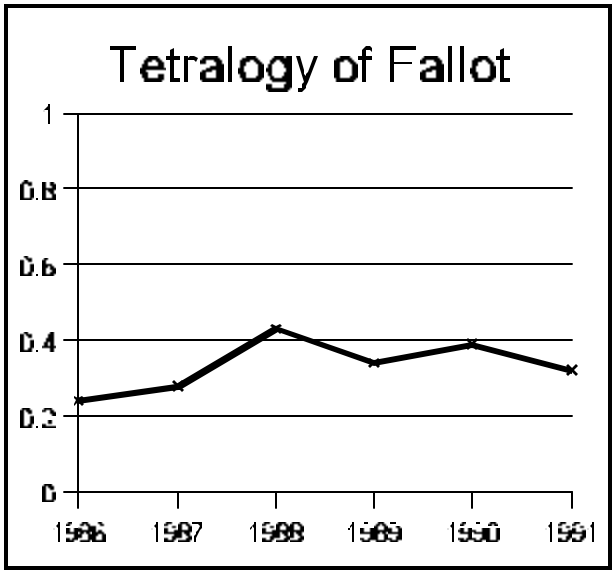
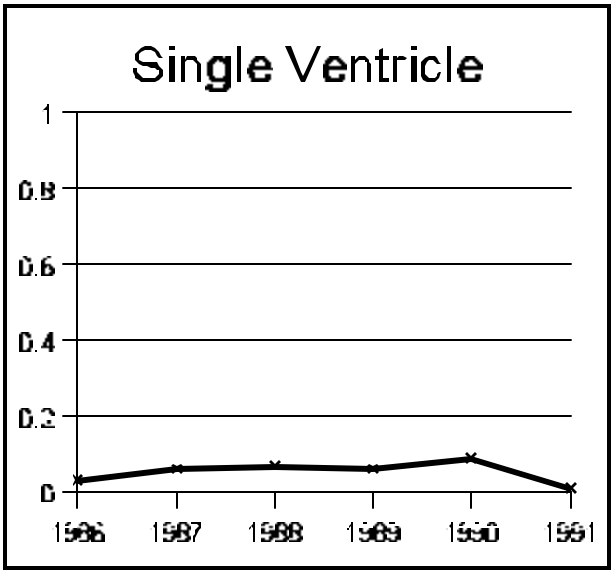


Figure 2. Trends of Selected Congenital Anomalies: Incident Rates
per 1,000 Live Births & Fetal Deaths, Arizona







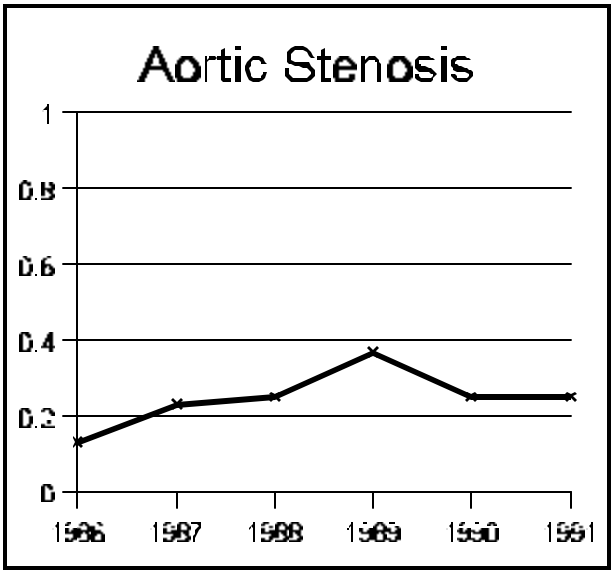
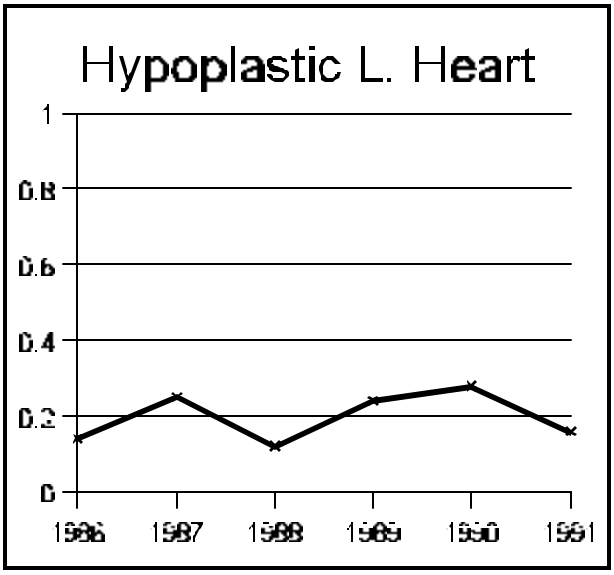
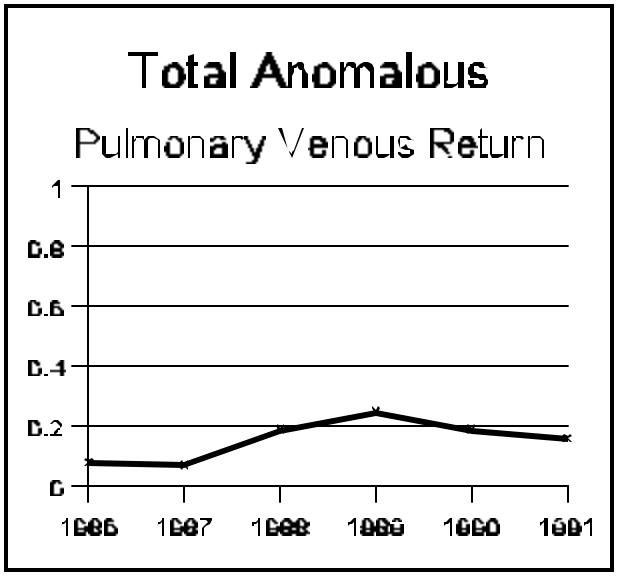
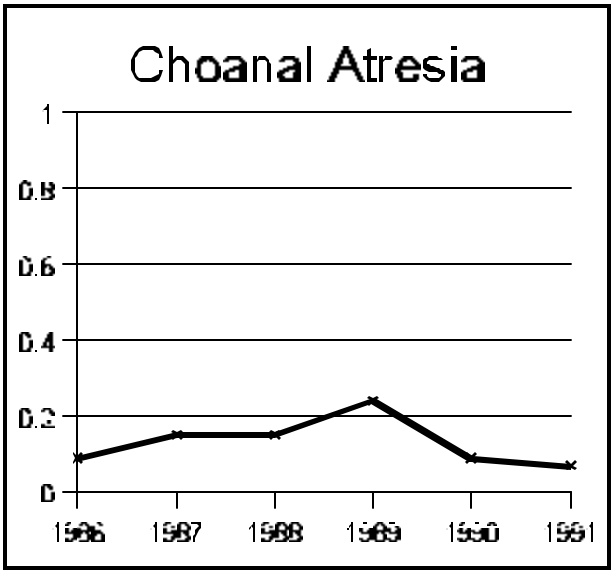
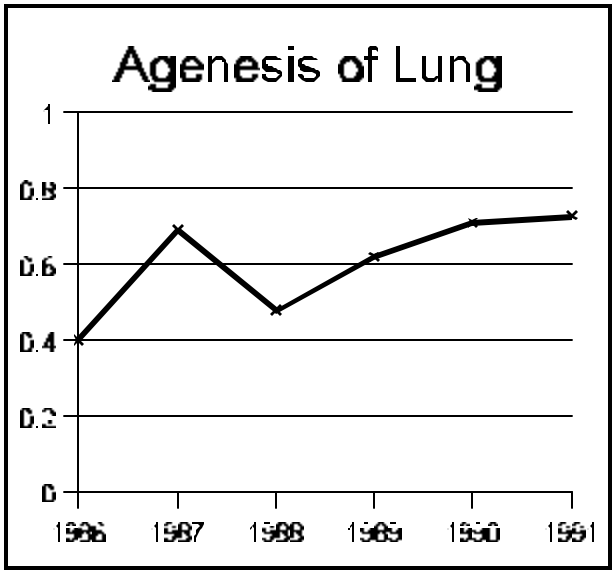


Figure 2. Trends of Selected Congenital Anomalies: Incident Rates per 1,000 Live Births & Fetal Deaths, Arizona









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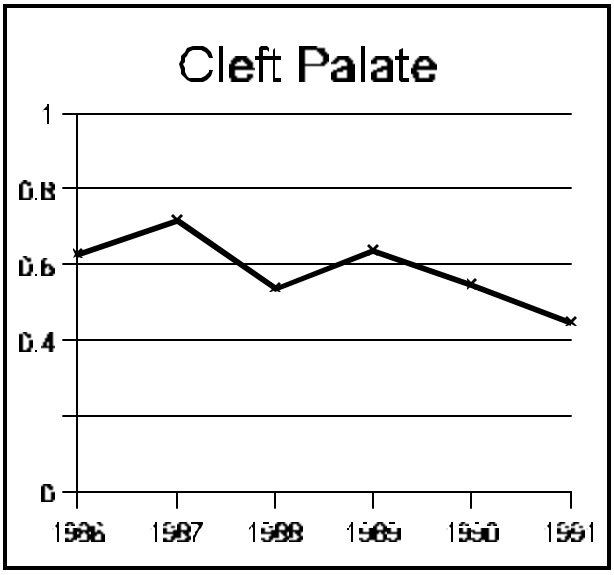
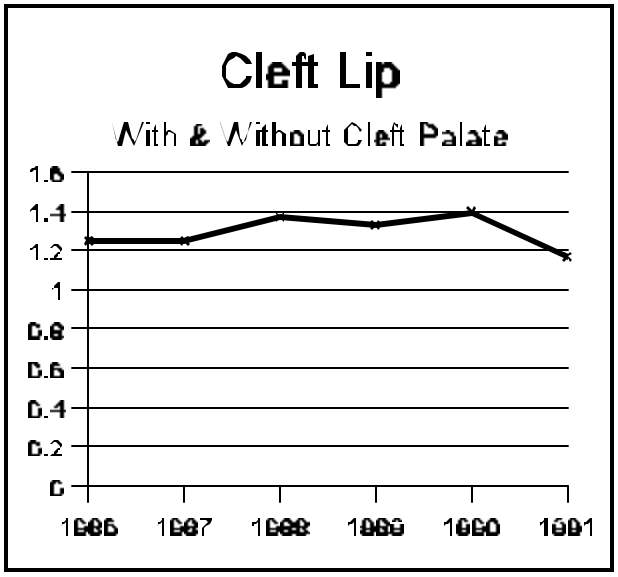
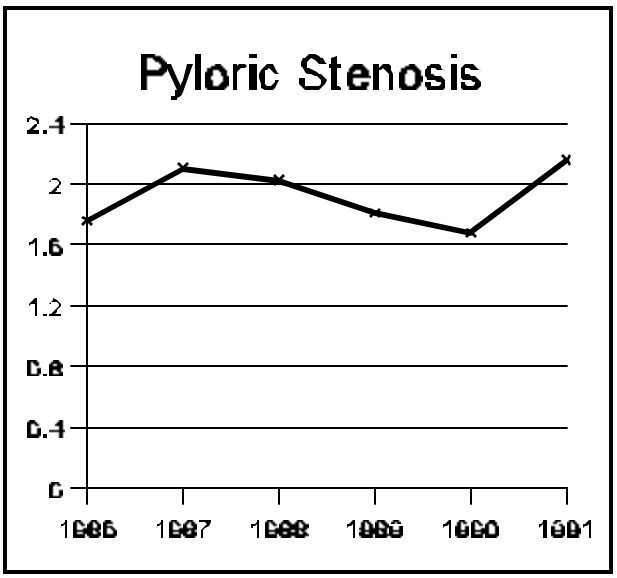
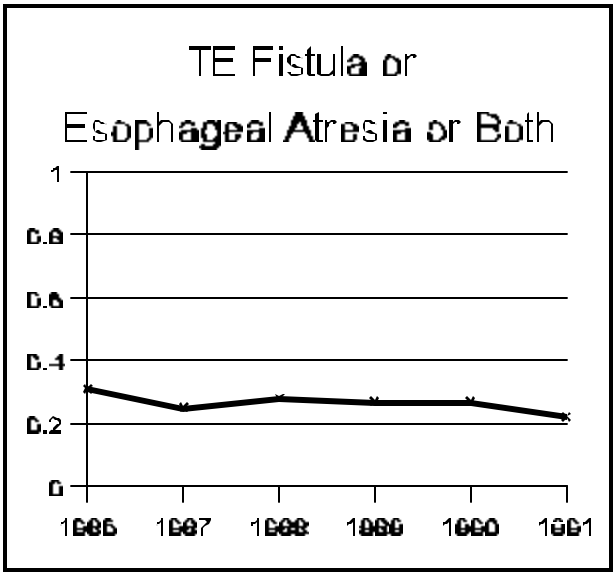
per 1,000 Live Births & Fetal Deaths, Arizona









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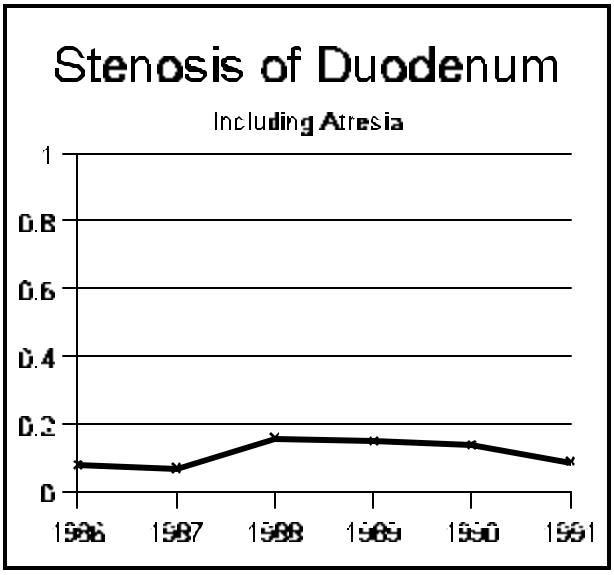
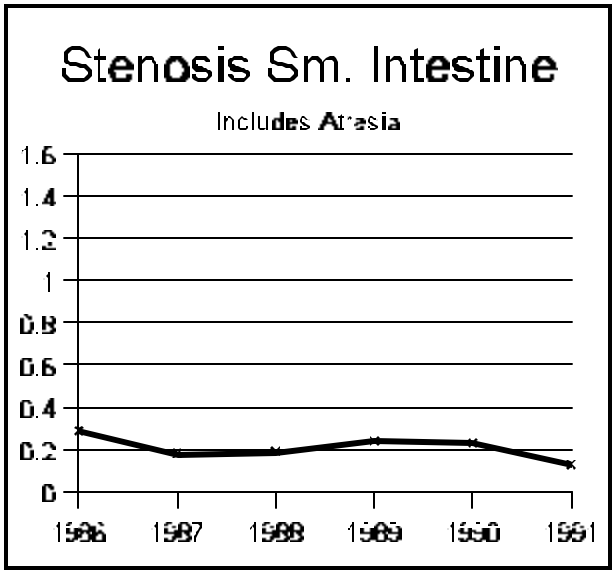
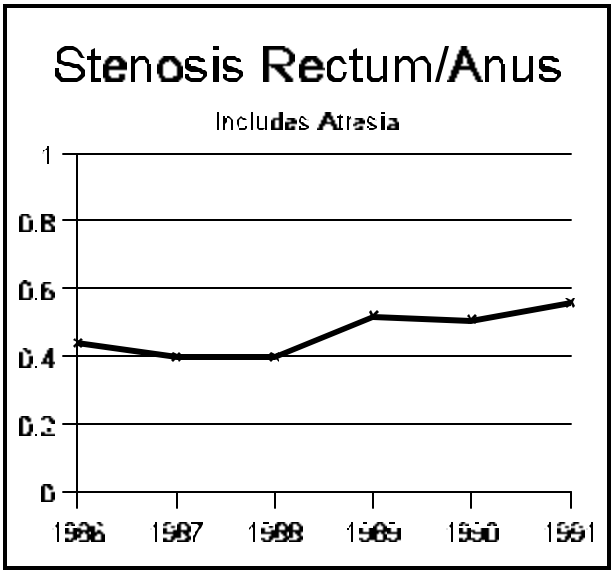






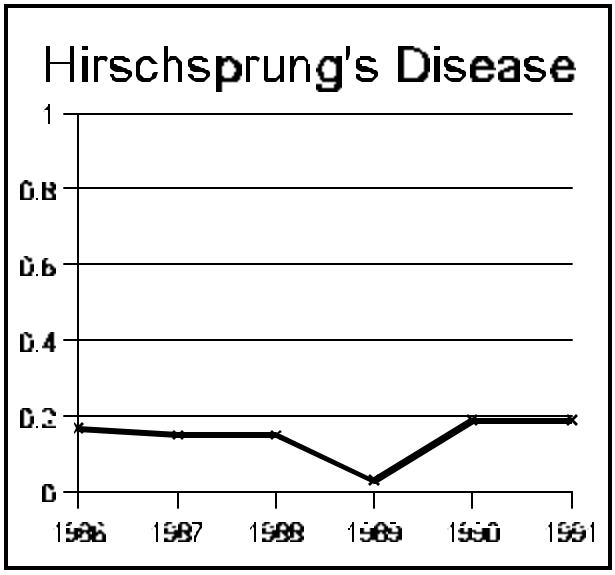
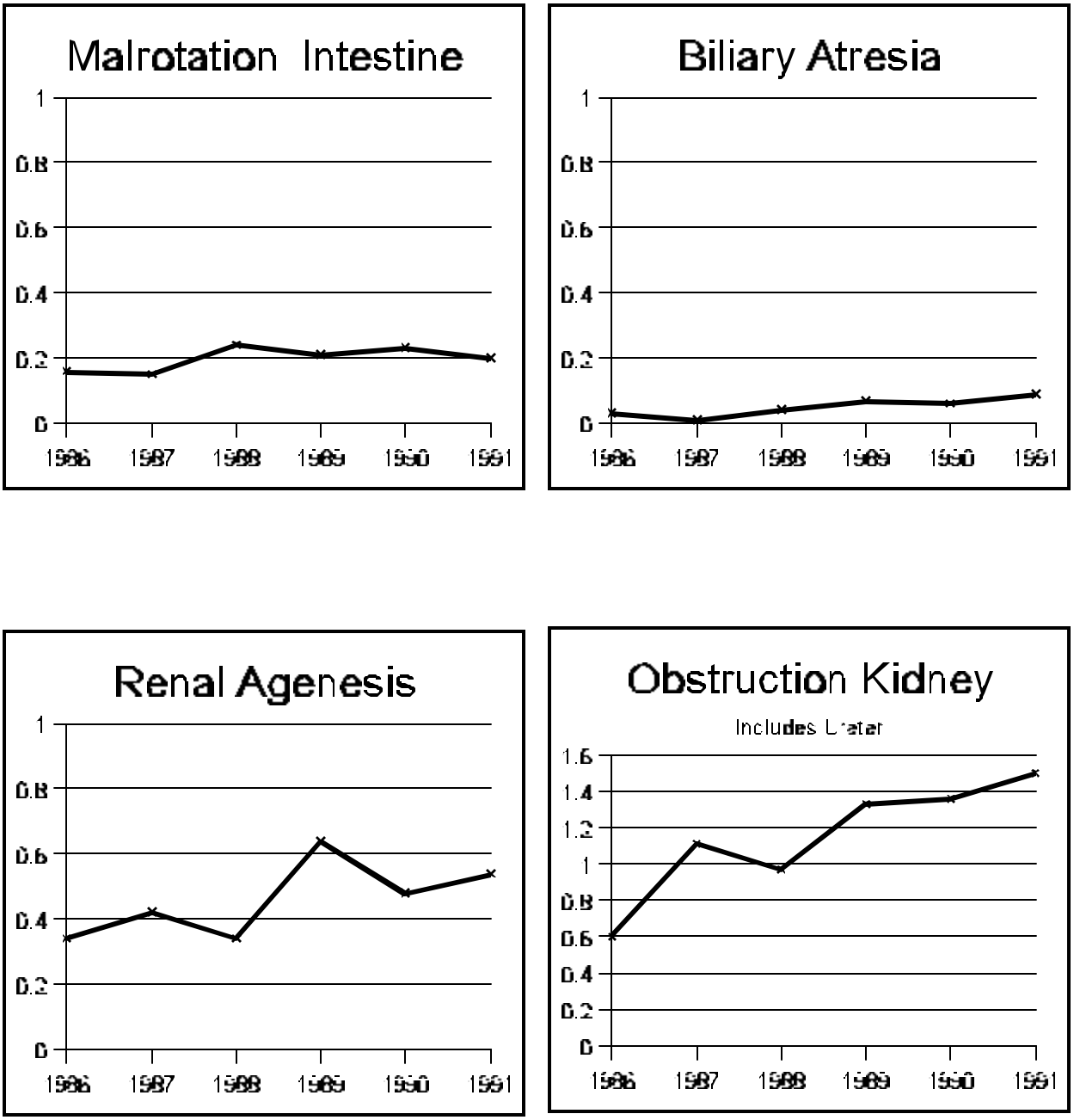


Figure 2. Trends of Selected Congenital Anomalies: Incident Rates per 1,000 Live Births & Fetal Deaths, Arizona



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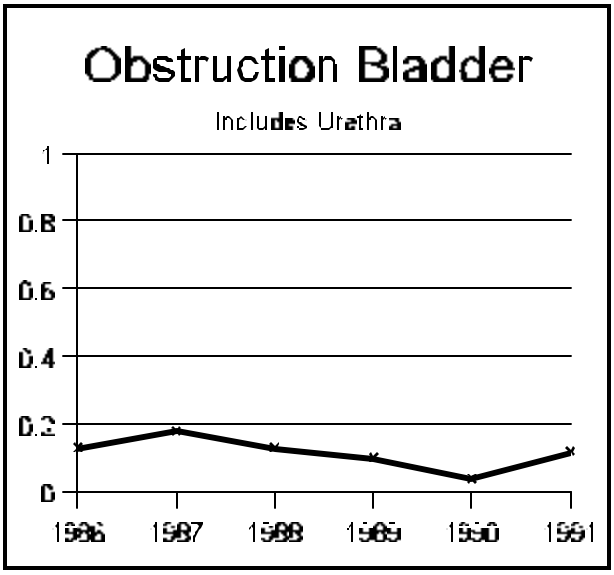
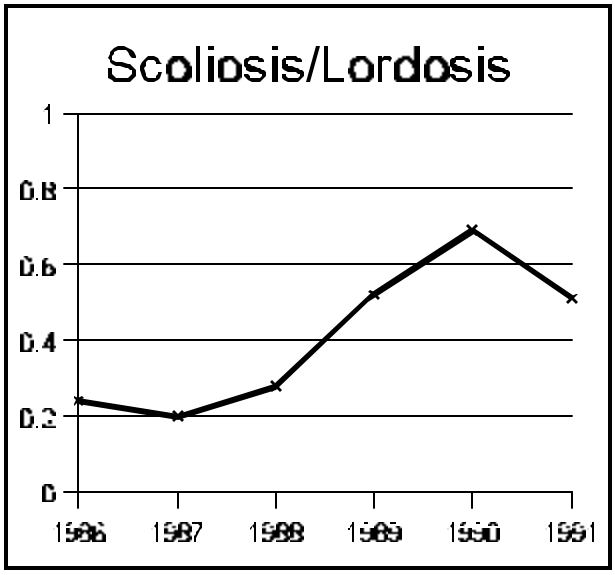
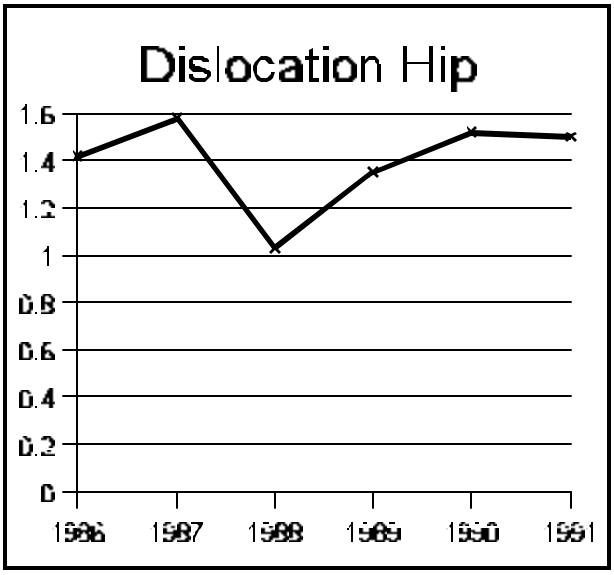






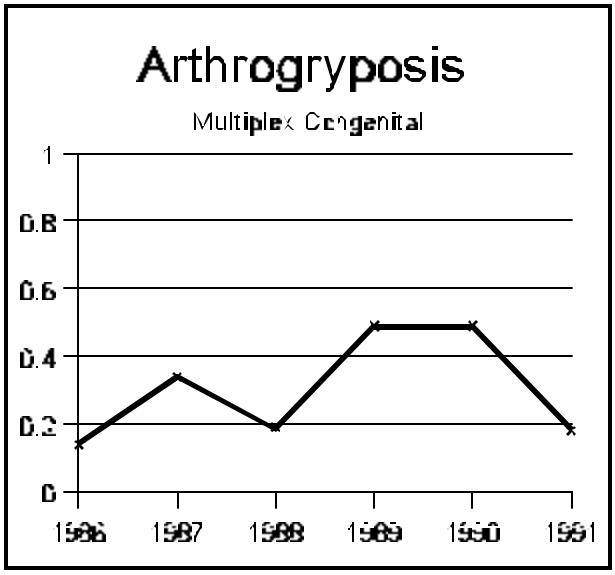
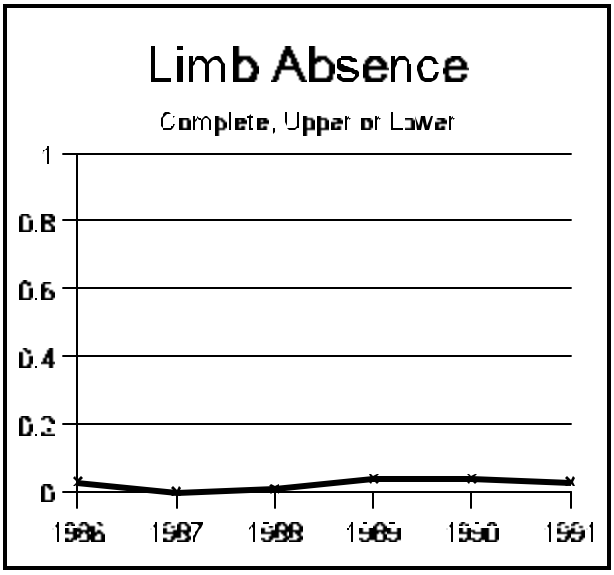
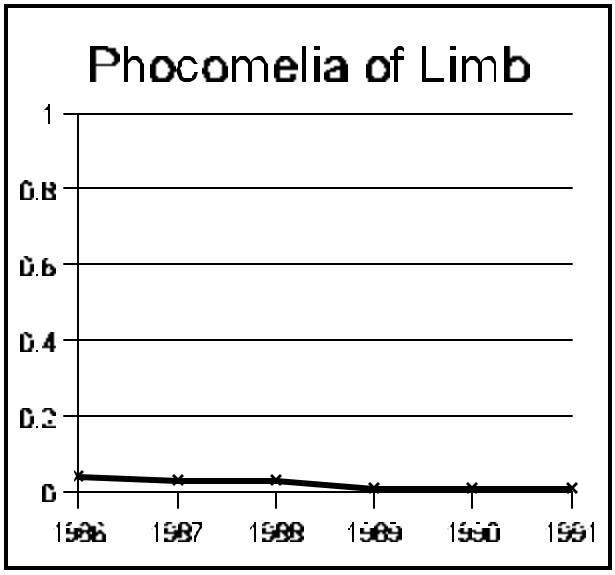
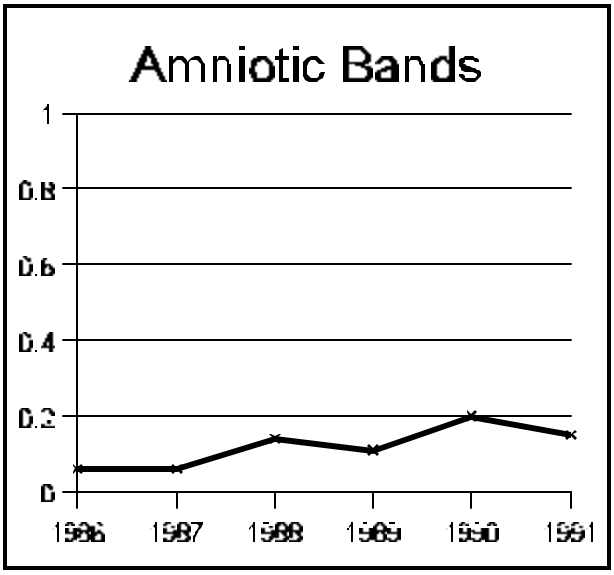


Figure 2. Trends of Selected Congenital Anomalies: Incident Rates per 1,000 Live Births & Fetal Deaths, Arizona







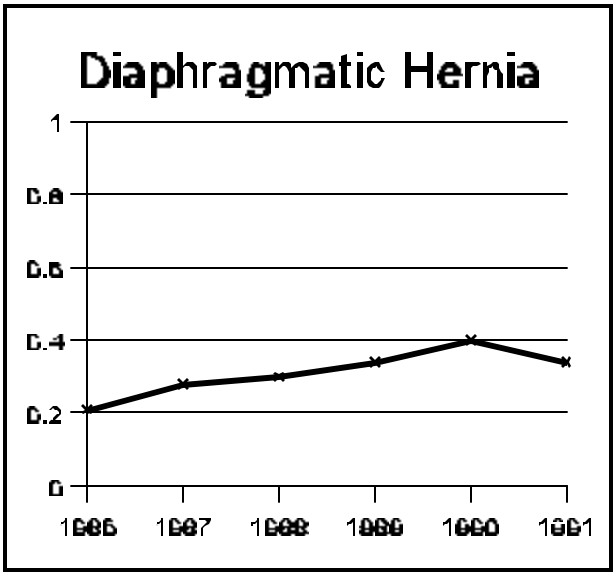


Figure 2. Trends of Selected Congenital Anomalies: Incident Rates per 1,000 Live Births & Fetal Deaths, Arizona

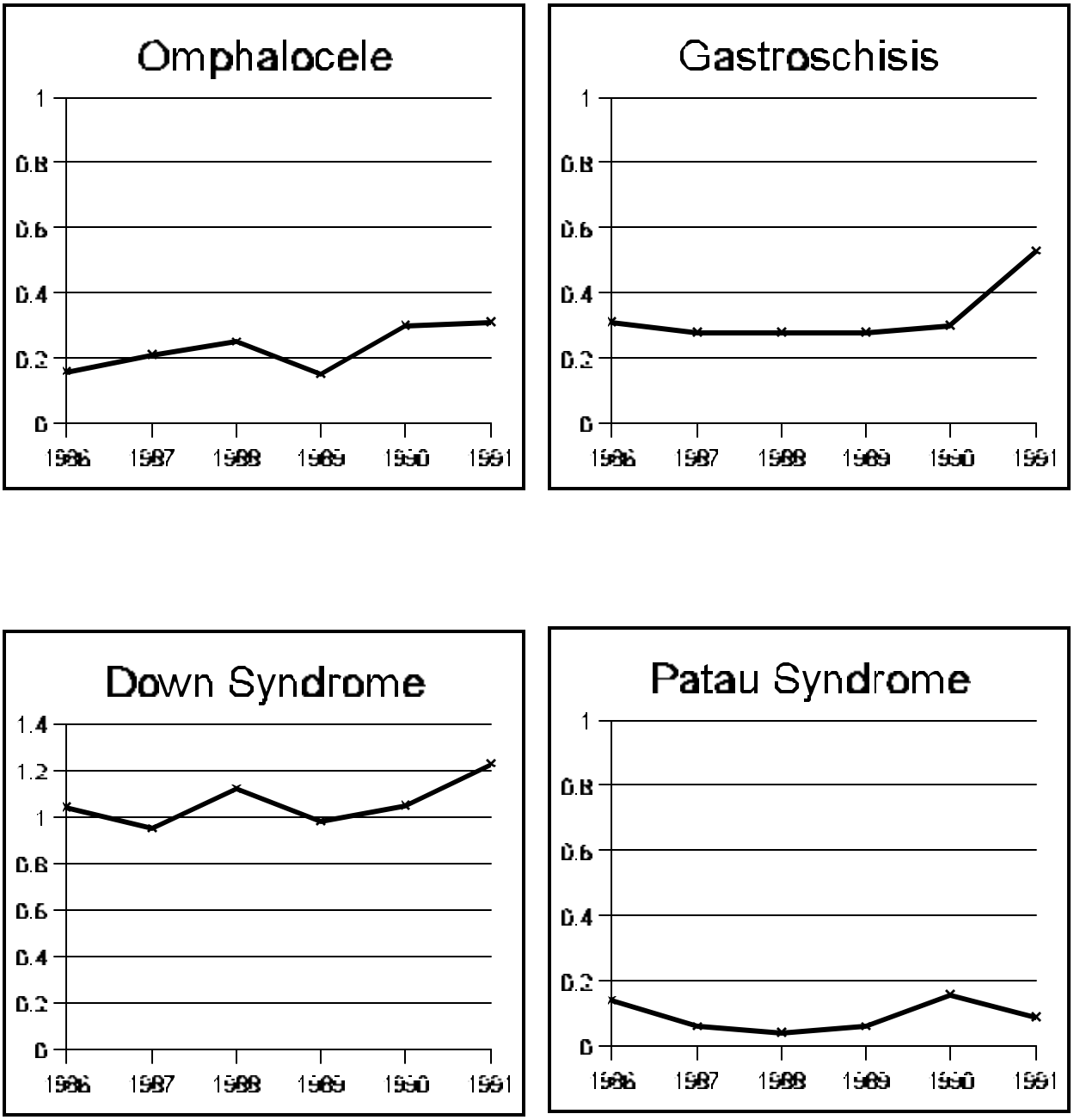
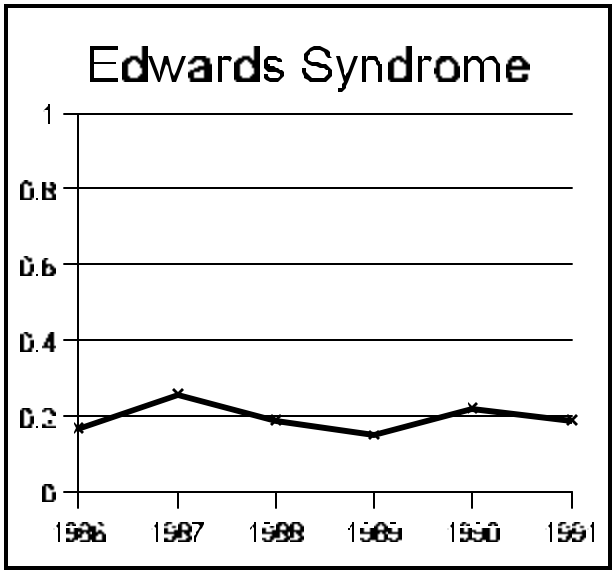
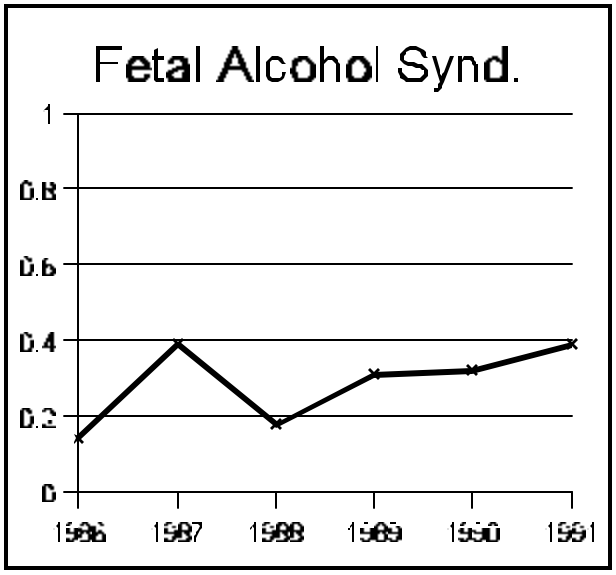
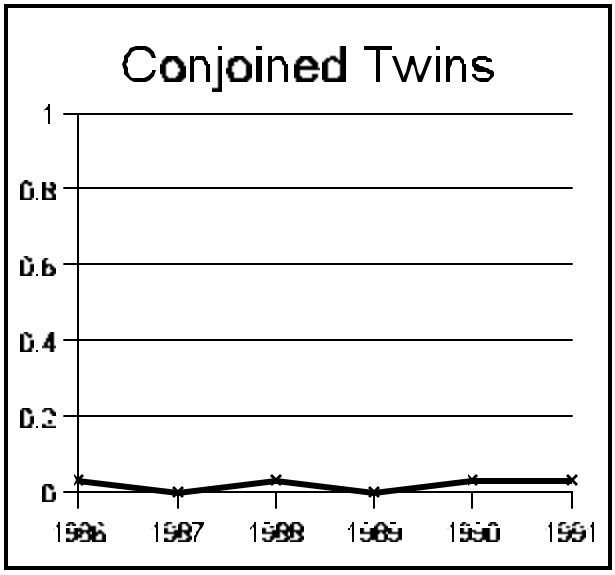


Figure 2. Trends of Selected Congenital Anomalies: Incident Rates per 1,000 Live Births & Fetal Deaths, Arizona







RACE/ETHNICITY

Race and ethnicity categories were determined from the mother's race and Hispanic origin portions of the child's birth certificate. The Hispanic category consists of mothers who answered 'White' to race and 'Hispanic' to the Hispanic origin question. The remaining race categories are Black, Native American, and Other.

Spina Bifida was the most common neural tube defect (NTD) among all races. Rates of Spina Bifida were highest among Hispanics (Figure 3). Many studies have documented that Hispanics have higher rates of Spina Bifida compared to Whites. In contrast, rates of Anencephaly were higher for Whites relative to Hispanics. The literature also suggests that Blacks experience lower rates of Spina Bifida and Anencephaly compared to Whites; however, we had to limit our rate comparisons of NTDs to White and Hispanic due to small number of cases occurring among other races.

The incidence of abdominal wall defects was highest among Hispanics compared to other races (Figure 4). Examining specific defects, we found that Gastroschisis rates were higher among Hispanics, relative to Whites. However, the reverse was observed for Omphalocele rates, for which Whites had the highest rate. Again, rate comparisons among other races was not possible due to small numbers.

Down Syndrome (Trisomy 21) rates were highest among Native Americans followed by Hispanics and Whites (figure 5).

For Native Americans and Blacks, microcephaly was the most frequently occurring birth defect, while for Whites and Hispanics it was pyloric stenosis (Figures 6 and 7).

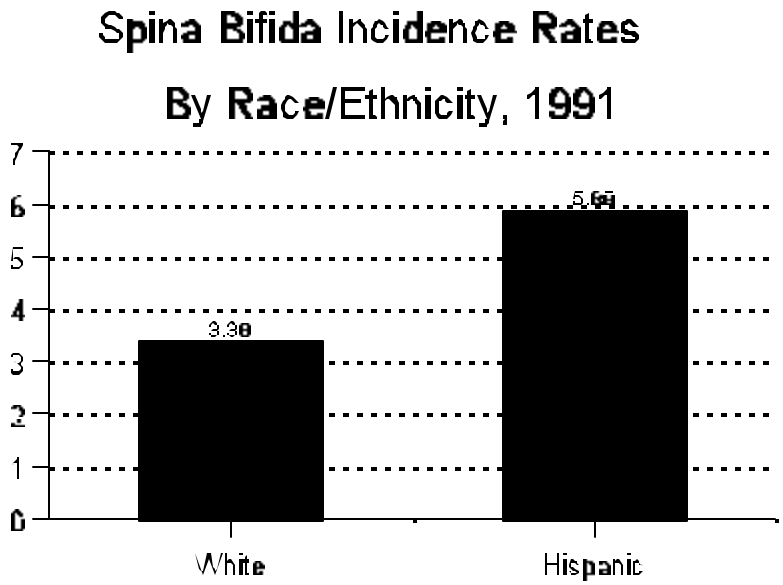


Figure 3. Spina Bifida Incidence Rates Per 10,000 Live Births and Fetal Deaths By Race/Ethnicity, 1991

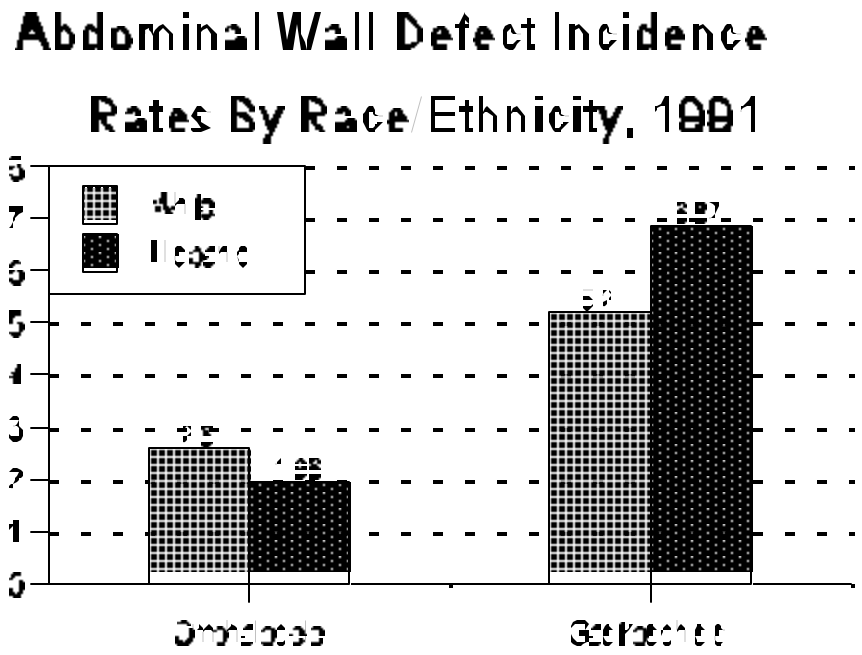


Figure 4. Abdominal Wall Defect Incidence Rates Per 10,000 Live Births and Fetal Deaths By Race/Ethnicity, 1991

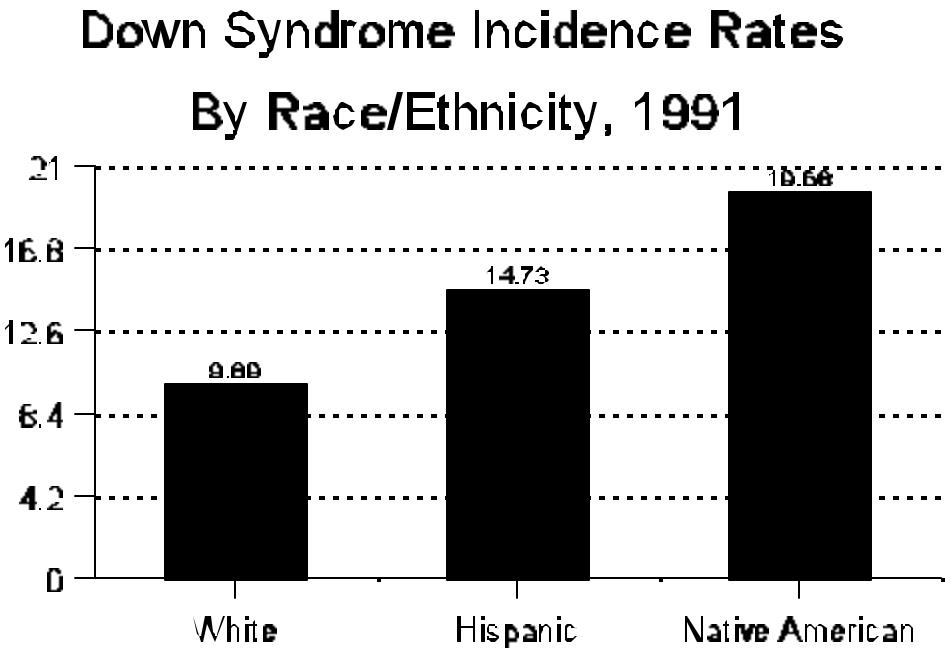


Figure 5. Down Syndrome Incidence Rates Per 10,000 Live Births and Fetal Deaths By Race/Ethnicity, 1991

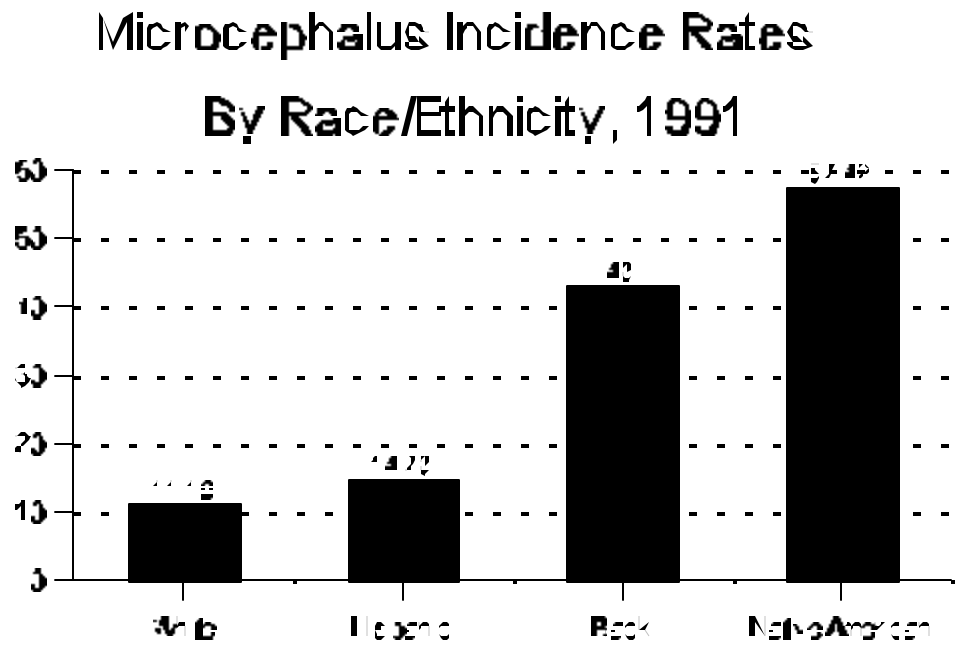


Figure 6. Microcephalus Incidence Rates Per 10,000 Live Births and Fetal Deaths By Race/Ethnicity, 1991

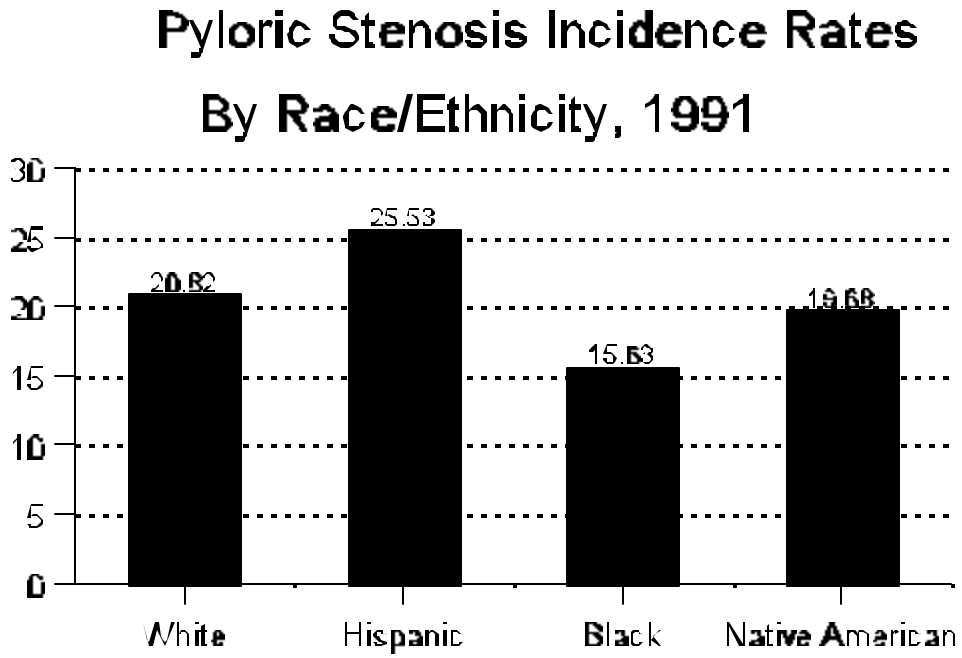


Figure 7. Pyloric Stenosis Incidence Rates Per 10,000 Live Births and Fetal Deaths By Race/Ethnicity, 1991

MATERNAL AGE

Maternal age was divided into five age groups. Observed rates of the "47 selected" congenital anomalies were highest among women 35 years of age and older, followed by the less than 20 age group (Figure 8). Down syndrome (Trisomy 21) rates increased with maternal age (Figure 9). In contrast, rates for gastroschisis decreased as maternal age increased (Figure 10).

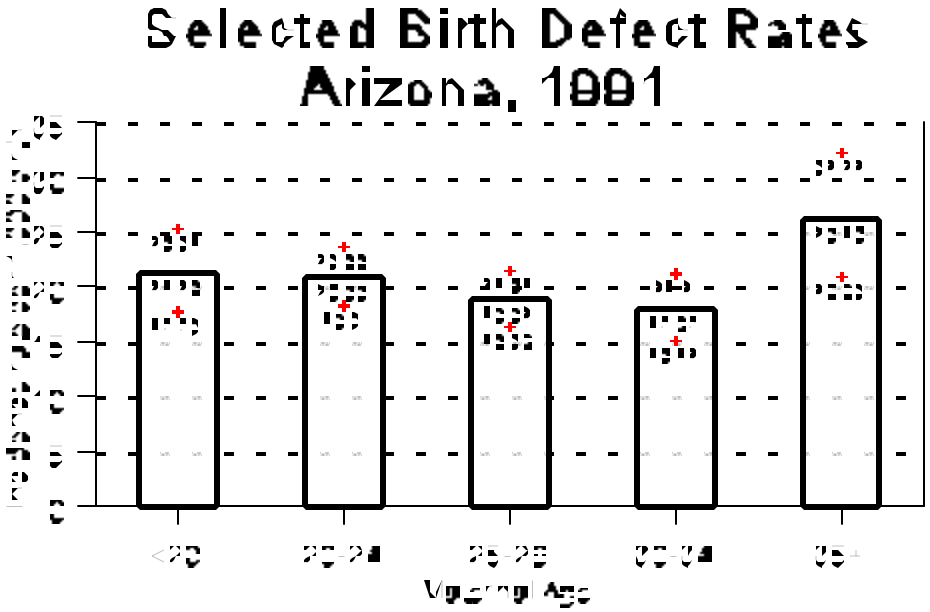


Figure 8. Incidence Rates per 1,000 Live Births and Fetal Deaths for the 47 Selected Defects Listed on Table 1. The [sign indicates the 99% confidence bounds.

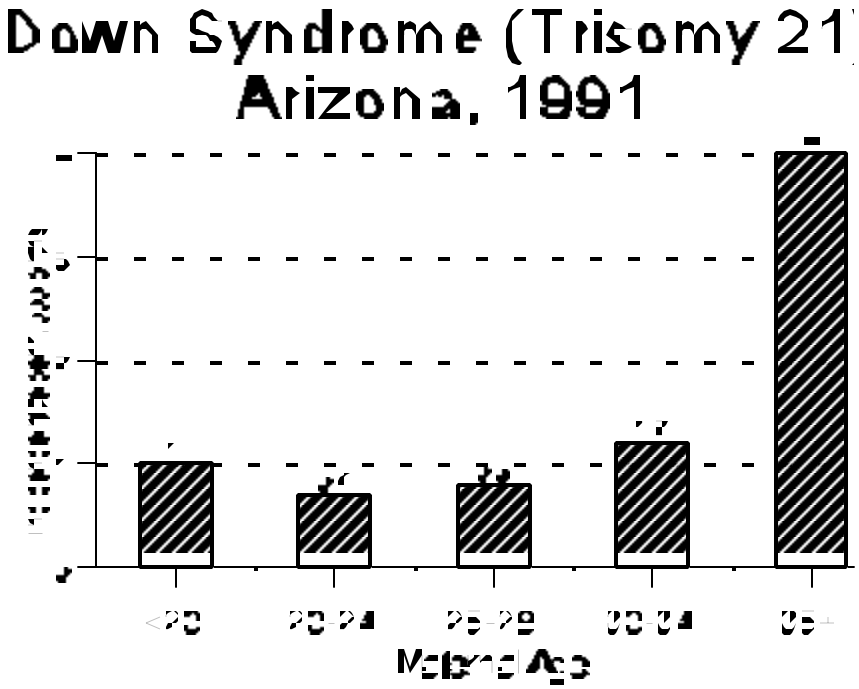


Figure 9. Down Syndrome (Trisomy 21) Rates Per 1,000 Live births and Fetal Deaths by Maternal Age.

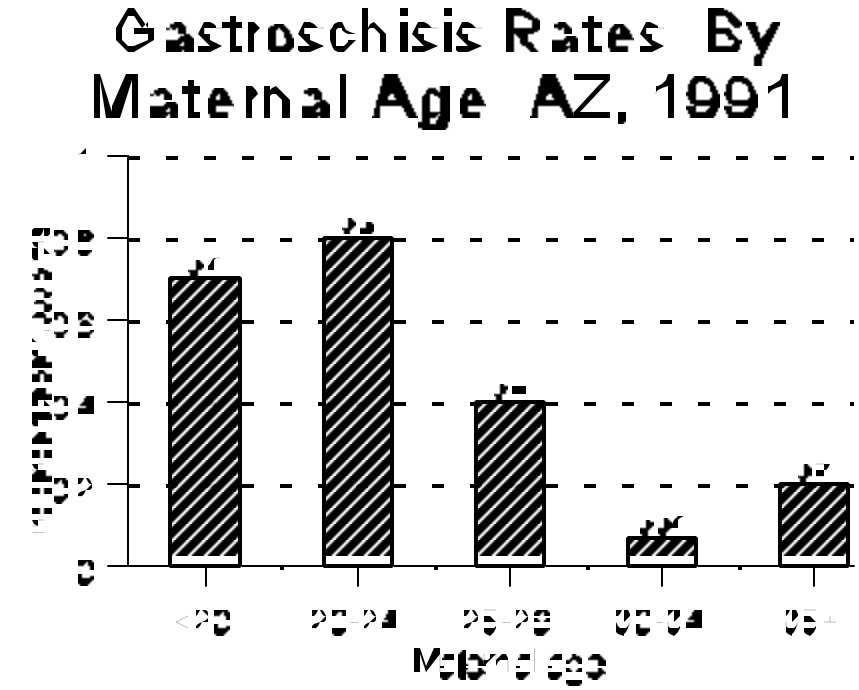


Figure 10. Gastroschisis Rates Per 1,000 Live births and Fetal Deaths by Maternal Age.

COUNTY PROFILES

Using County Data

The Arizona Birth Defect Monitoring Program (ABDMP) collects birth defect information from all of Arizona's 15 counties. This is the first year that the ABDMP has presented county specific data on birth defects. Until this time there has not been a large enough aggregate of data to derive reliable statistical measures at the county level.

Dealing With Small Numbers

Analysis of county data is difficult because of normal fluctuations in rates seen in small populations. When dealing with small numbers, it is normal to see fluctuations over time. With rate fluctuations we may see the appearance of birth defects clusters, most often this is a statistical anomaly. In the rare case that a cluster results from a teratogen a dramatic increase on the scale of 10-fold or greater is usually seen.[5] Another concern with small numbers is protecting a person and their family's confidentiality. Thus, all county level data are aggregated. Incidence rates and confidence intervals will only be presented when there are 10 or more cases.

Birth Defects by County

The following tables present birth defects by county of mothers' residence. Table 4 shows the total number of 47 selected congenital anomalies for each Arizona county. Table 5 examines selected anomalies by race and county. Cases were aggregated for the years 1986 through 1991 to provide large enough numbers for analysis. Gila county had the highest rate of congenital anomalies, whereas Greenlee and La Paz counties had the lowest reported birth defects rates. However, there was no significant difference in overall rates between any of Arizona's 15 counties. Five sentinel defects (chromosomal defects, oral clefts, heart defects, abdominal wall defects, and neural tube defects) were examined by county (Tables 6-10).

Table 4
Selected Birth Defect Incidence Rates by County 1986-1991
Rates Per 1,000 Live Births

COUNTY	CASES 1986-1991	RATE	99% CONFIDENCE INTERVAL
Apache	202	18.67	15.46-22.34
Cochise	159	16.13	13.02-19.73
Coconino	199	16.93	13.99-20.28
Gila	105	26.97	20.66-34.54
Graham	43	16.72	10.87-24.50
Greenlee	5	-	-
Maricopa	4,048	17.63	16.93-18-36
Mohave	98	12.46	09.45-16.09
Navajo	236	20.22	16.98-23.87
Pima	1159	17.42	16.13-18.78
Pinal	234	18.54	15.56-21.90
Santa Cruz	64	16.10	11.38-22.06
Yavapai	133	18.28	14.45-22.78
Yuma	245	18.34	15.46-21.58
La Paz	17	14.29	06.91-25.91

47 selected birth defects (see Table 1); - =Insufficient cases for rate and confidence interval calculations.

Table 5 – Selected Birth Defects by Race/Ethnicity by County, 1986-1991
Incidence Rates Per 1,000 live births

COUNTY	WHITE	HISPANIC	BLACK	NATIVE AMERICAN	OTHER
	Rate 99% C.I.	Rate 99% C.I.	Rate 99% C.I.	Rate 99% C.I.	Rate 99% C.I.
Apache	8.64 3.37-17.93	- -	- -	20.54 16.88-24.73	- -
Cochise	16.64 12.26-22.04	15.80 11.00-21.93	18.59 8.50-34.95	- -	- -
Coconino	11.71 8.06-16.39	15.99 8.09-28.14	- -	21.66 16.88-27.37	- -
Gila	13.52 7.45-22.42	17.84 8.16-33.54	- -	50.97 35.92-70.01	- -
Graham	14.63 7.40-25.73	- -	- -	40.20 18.94-74.19	- -
Greenlee	- -	- -	- -	- -	- -
Maricopa	16.79 15.93-17.68	19.04 17.62-20.55	16.14 13.27-19.42	26.68 21.80-32.29	12.12 8.04-17.48
Mohave	12.39 9.18-16.32	- -	- -	- -	- -
Navajo	14.18 9.31-20.60	24.67 11.93-44.75	- -	22.37 18.22-27.16	- -
Pima	15.80 14.11-17.62	18.05 15.94-20.37	22.08 15.56-30.33	26.97 19.70-35.96	9.78 4.00-19.71
Pinal	13.80 10.06-18.44	19.14 14.31-25.03	- -	35.19 24.95-48.11	- -
Santa Cruz	- -	16.74 11.69-23.17	- -	- -	- -
Yavapai	17.95 13.84-22.85	20.434 10.12-36.48	- -	- -	- -
Yuma	13.54 9.59-18.50	20.47 16.52-25.06	- -	76.09 33.69-146.09	- -
La Paz	- -	- -	- -	- -	- -

- =Insufficient cases for rate and confidence interval calculations.

SENTINEL DEFECTS

Tables 6-10 look at the following sentinel defects: chromosomal defects, oral clefts, neural tube defects, abdominal wall defects, and heart defects. These defects were chosen because of their significant public health impact.

Chromosomal Defects

In this section (Table 6) of the report chromosomal defects refers to Down Syndrome, Patau syndrome, and Edwards syndrome. Chromosomal abnormalities include either missing or extra genetic components that result in various levels of abnormal physical features, structural defects, and mental retardation. The most common chromosomal defects is Down Syndrome. We also know that the risk of a trisomy affected pregnancy increases with maternal age; however, this risk is still relatively low. Recent research also suggests that about 20% of instances of Down Syndrome, are paternal in origin.

Oral Clefts

Table 7 presents information on cleft lip and cleft palate. Cleft palate is a failure of the palate to fuse properly, forming a grooved fissure in the roof of the mouth. Cleft lip is a failure of the maxillary and median nasal processes to fuse, forming a fissure in the lip. Babies born with oral clefts require corrective surgery, and may have feeding problems. Mothers who smoke 20 or more cigarettes a day are more than twice as likely to have a baby born with cleft lip and/or cleft palate.[6]

Heart Defects

This category includes truncus Arteriosus, transposition of great vessels, Tetralogy of Fallot, single ventricle, aortic stenosis, hypoplastic left heart, and total anomalous pulmonary venous (Table 10).

Abdominal Wall Defects

This category includes omphalocele and gastroschisis (Table 9). Gastroschisis is a congenital opening of the abdominal wall, often with protrusion of the intestines. Omphalocele is a membrane-covered protrusion of an abdominal organ through the abdominal wall at the umbilicus. According to a recent study, young mothers are 4 times as likely as women in their late 20s to have a child with gastroschisis.[7] Other risk factors for gastroschisis are maternal use of cocaine, aspirin, amphetamines, and exposure to solvents.

Neural Tube Defects

Anencephaly, spina bifida, and encephalocele make up the neural tube defects (NTDs) presented in Table 8. The two major NTDs are anencephaly and spina bifida. Anencephaly is an absence of the skull, with cerebral hemispheres reduced or completely missing. Spina bifida is a defective closure of the bony encasement of the spinal cord, through which the cord and meninges may or may not protrude. Women who take multivitamins and/or eat a diet rich in folate can significantly reduce their risk of an NTD affected pregnancy.

FOLATE FACTS
WHAT IS FOLATE?
Folate is a B vitamin. It is also called folic acid or folacin. Folate helps your body form red blood cells. It also helps a baby's spine and brain develop before it is born.
WHY IS FOLATE IMPORTANT?
You need folate in your body before you get pregnant and especially in the first months of pregnancy. This greatly reduces your chance of having a baby with a birth defect of the brain or spine. Folate also reduces you risk of developing heart disease, and possibly certain forms of cancer.
HOW DO YOU GET FOLATE?
Eat folate-rich foods and take a multivitamin daily. Some foods rich in folate include: orange juice, dried beans, fortified breakfast cereals, broccoli, cauliflower, and corn.
HOW MUCH FOLATE?
The United States Public Health Service now recommends all women of childbearing age take a supplement containing 0.4 milligrams of folic acid daily.

Table 6
Chromosomal Defects - Rates by County 1986-1991
Incidence Rate 1,000 Live Births

COUNTY	CASES 1986-1991	RATE	99% CONFIDENCE INTERVAL
Apache	18	1.66	0.82-2.97
Cochise	16	1.62	0.76-2.99
Coconino	13	1.11	0.47-2.17
Gila	10	2.57	0.95-5.51
Graham	5	-	-
Greenlee	0	-	-
Maricopa	284	1.24	1.06-1.44
Mohave	7	-	-
Navajo	19	1.63	0.82-2.86
Pima	80	1.20	0.88-1.59
Pinal	17	1.35	0.65-2.44
Santa Cruz	6	-	-
Yavapai	9	-	-
Yuma	15	1.12	0.51-2.11
La Paz	3	-	-

Chromosomal defects include three-digit codes R01, R02, R03 (see Table 1);
- =Insufficient cases for rate and confidence interval calculations.

Table 7
Oral Clefts - Rates by County 1986-1991
Incidence Rates Per 1,000 Live Births

COUNTY	CASES 1986-1991	RATE	99% CONFIDENCE INTERVAL
Apache	36	3.33	2.07-5.05
Cochise	25	2.54	1.42-4.16
Coconino	27	2.30	1.32-3.70
Gila	13	3.34	1.43-6.56
Graham	8	-	-
Greenlee	0	-	-
Maricopa	370	1.61	1.40-1.84
Mohave	10	1.27	0.47-2.73
Navajo	31	2.66	1.59-4.15
Pima	112	1.68	1.30-2.14
Pinal	26	2.06	1.17-3.35
Santa Cruz	4	-	-
Yavapai	18	2.47	1.22-4.42
Yuma	34	2.54	1.56-3.90
La Paz	2	-	-

Oral Clefts include three-digit codes F01 & F02 (see Table 1); - =Insufficient cases for rate and confidence interval calculations.

Table 8
Neural Tube Defects - Rates by County 1986-1991
Incidence Rates Per 1,000 Live Births

COUNTY	CASES 1986-1991	RATE	99% CONFIDENCE INTERVAL
Apache	7	-	-
Cochise	6	-	-
Coconino	5	-	-
Gila	3	-	-
Graham	2	-	-
Greenlee	0	-	-
Maricopa	184	0.80	0.66-0.97
Mohave	6	-	-
Navajo	12	1.03	0.42-2.07
Pima	46	0.69	0.46-1.00
Pinal	9	-	-
Santa Cruz	6	-	-
Yavapai	7	-	-
Yuma	13	0.97	0.42-1.91
La Paz	3	-	-

Neural Tube defects include three-digit codes A01, A02, A03 & A13.
(see Table 1); - =Insufficient cases for rate and confidence interval calculations.

Table 9
Abdominal Wall Defects - Rates by County 1986-1991
Incidence Rates Per 1,000 Live Births

COUNTY	CASES 1986-1991	RATE	99% CONFIDENCE INTERVAL
Apache	2	-	-
Cochise	3	-	-
Coconino	3	-	-
Gila	3	-	-
Graham	1	-	-
Greenlee	0	-	-
Maricopa	109	0.47	0.37-0.61
Mohave	7	-	-
Navajo	7	-	-
Pima	46	0.69	0.46-1.00
Pinal	14	1.11	0.49-2.13
Santa Cruz	2	-	-
Yavapai	7	-	-
Yuma	7	-	-
La Paz	0	-	-

Abdominal Wall defects include three-digit codes N02 & N04 (see Table 1); - =Insufficient cases for rate and confidence interval calculations.

Table 10
Heart Defects - Rates by County 1986-1991
Incidence Rates Per 1,000 Live Births

COUNTY	CASES 1986-1991	RATE	99% CONFIDENCE INTERVAL
Apache	12	1.11	0.45-2.24
Cochise	14	1.42	0.63-2.73
Coconino	12	1.02	0.42-2.06
Gila	10	2.57	0.95-5.51
Graham	2	-	-
Greenlee	1	-	-
Maricopa	346	1.51	1.31-1.73
Mohave	7	-	-
Navajo	22	1.88	1.01-3.19
Pima	110	1.65	1.27-2.11
Pinal	18	1.43	0.71-2.55
Santa Cruz	8	-	-
Yavapai	15	2.06	0.94-3.88
Yuma	22	1.65	0.88-2.79
La Paz	0	-	-

Heart defects include three-digit codes D01, D02, D03, D04, D51, D52 & D53 (see Table 1); - =Insufficient cases for rate and confidence interval calculations.

APPENDIX 1

Conditions Included in the Figures

A general listing of all conditions used to establish the rates shown in the figures in this report is shown below. Some specific inclusions and exclusions are not listed. As mentioned above, ABDMP collects data on hundreds of conditions or variations of conditions. The conditions listed below include over 99% of all cases reported through ABDMP.

BPA 3-Digit Code*	General Code Descriptor
740 - 759	"Congenital Anomalies" Including but not limited to:
740	Anencephaly and similar anomalies
741	Spina Bifida
742	Other Anomalies of the Nervous System
743	Anomalies of the eye
744	Anomalies of the ear, face, and neck
745	Certain anomalies of the heart
746	Other anomalies of the heart
747	Anomalies of the circulatory system
748	Anomalies of the respiratory system
749	Cleft palate and cleft lip
750	Other anomalies of the upper alimentary tract
751	Anomalies of the digestive system
752	Anomalies of the genital organs
753	Anomalies of the urinary system
754	Certain musculoskeletal deformities
755	Other anomalies of limbs
756	Other musculoskeletal anomalies
757	Congenital anomalies of the integument
758	Chromosomal anomalies
759	Other and unspecified anomalies
ICD-9-CM Code**	
090.00-090.9	Congenital syphilis
658.80-658.83	Amniotic bands
760.71	Fetal alcohol syndrome
771.0-771.2	Congenital infections including but not limited to: rubella, cytomegalovirus, toxoplasmosis, and herpes simplex.

* British Pediatric Association Classification of Diseases

** International Classification of Disease - 9th Edition, Clinical Modification

APPENDIX 2

Conditions Shown in the Tables

A listing of the conditions analyzed in the Tables contained in this report is shown below.

The 47 conditions listed here can be described almost completely by codes created by the Centers for Disease Control's Metropolitan Atlanta Congenital Defects Program (MACDP). These codes are listed in the left below, with exceptions noted. On the right below are the corresponding British Pediatric Association (BPA) Classification of Diseases codes.

In the Tables, a case is listed only once in each MACDP code category, even when it has more than one diagnosis from the category.

MACDP Code	Condition	BPA Code		
<u>CENTRAL NERVOUS SYSTEM</u>				
A01	Anencephaly	740.00	740.02	740.03
		740.08	740.10	740.20
		740.21	740.29	740.20
A02	Spina Bifida without Hydrocephaly	741.00	741.01	741.02
		741.03	741.04	741.05
		741.06	741.07	741.08
		741.09	741.87	
A03	Spina Bifida with hydrocephaly	741.90	741.91	741.92
		741.93	741.94	741.98
		741.99		
A13	Encephalocele	742.00	742.08	742.09
		742.085	742.086	
A15	Hydrocephaly	742.30	742.31	742.38
		742.39		
A16	Microcephalus	742.10		

EYE AND EAR				
B03	Glaucoma	743.20	743.21	743.22
B04	Cataract	743.32		
B51*	Anophthalmia	743.00		
B52*	Microphthalmia	743.10		
B54*	Ear anomaly with hearing loss	744.00	744.01	744.02
		744.03	744.09	744.21
CARDIAC				
D01	Truncus Arteriosus	745.00	745.01	
D02	Transposition of great vessels	745.10	745.11	745.12
		745.18	745.19	
D03	Tetralogy of Fallot	745.20	745.21	746.84
D04	Single ventricle	745.30		
D51*	Aortic stenosis	746.30	746.31	
D52*	Hypoplastic left heart	746.70		
D53*	Total anomalous pulmonary venous return	747.42		
RESPIRATORY				
E01	Choanal atresia	748.00		
E06	Agenesis of lung	748.50	748.51	
OROFACIAL - GASTRO-INTESTINAL				
F01	Cleft palate	749.00	749.01	749.02
		749.03	749.04	749.05
		749.06	749.07	749.09
F02	Cleft lip with or without cleft palate	749.10	749.11	749.12
		749.19	749.20	749.21
		749.22	749.29	

F08	Pyloric Stenosis	750.51		
F09	Tracheo-esophageal fistula or esophageal atresia	750.30	750.31	750.32
		750.33		
F14	Stenosis or atresia of duodenum	751.10		
F15	Other stenosis or atresia of small intestine	751.11	751.12	751.19
F16	Stenosis or atresia of rectum or anus	751.21	751.22	751.23
		751.24		
F17	Hirschsprung's Disease	751.30	751.31	751.32
		751.33		
F18	Malrotation of intestine	751.40	751.41	751.42
		751.49	751.495	
F21	Biliary atresia	751.65		
GENITO-URINARY				
H01	Renal agenesis	753.00	753.01	
H06	Obstruction of kidney or ureter	753.20	753.21	753.22
		753.29	753.40	753.42
H09	Bladder or urethra obstruction	753.600	753.61	753.62
		753.63		
MUSCULOSKELETAL				
J02	Curvature of spine (scoliosis or lordosis	754.20	754.21	754.22
J03	Dislocation of hip	754.30		
J11	Arthrogryposis multiplex congenital	755.80		
J51*	Complete absence of upper or lower limb	755.20	755.30	755.40
J52*	Phocomelia of Limb	755.21	755.31	755.41
K05	Amniotic bands	658.80		

N01	Diaphragamtic hernia	756.61	756.615-7	
N02	Omphalocele	756.70		
N03	Gastroschisis	756.71		
SYNDROMES				
R01	Down Syndrome	758.00	758.01	758.02
		758.03	758.04	758.09
R02	Patau Syndrome (Trisomy 13)	758.10	758.11	758.12
		758.13	758.19	
R03	Edwards Syndrome (Trisomy 18)	758.20	758.21	758.22
		758.23	758.29	758.295
S02	Fetal Alcohol Syndrome	760.71	760.718	
W03	Conjoined twins	759.40	759.41	759.42
		759.43	759.44	759.48
		759.49		

* Codes created by CBDMP

APPENDIX 3

PRECISION (of diagnosis) (Box 32 FORM 01)

Code

1	Not stated (For Mental Retardation and Cerebral Palsy Diagnoses ONLY - Form 03)
2	Probably not a birth defect ("Ruled out" included in this category), "NO"
3	"vs" (versus) or "or"
4	"Rule out" included in diagnosis (i.e., rule out anencephaly), "Doubtful," "Equivocal", "Questionable," "R/O"
5	"Suggestive of"
6	"Suspected," "suspicious"
7	"Possible," "may have," "could be," "felt to be," "Perhaps," "consider"
8	"Consistent with," "most likely"
9	"Compatible with," "like," "appears"
10	"Probable," "presume"
11	------
12	Precise diagnosis, "characteristic of"
13	Precise diagnosis with congestive heart failure or medicated with Digoxin, Drisdol, Chlorothiazide, Lasix, Lanoxin, Aldactone or diuretics (only for VSD, PDA, ASD, or Patent Foramen Ovale)

APPENDIX 4

Abbreviations

ABDMP	- Arizona Birth Defects Monitoring Program
ADHS	- Arizona Department of Health Services
BPA	- British Pediatric Association
CBDMP	- California Birth Defects Monitoring Program
CDC	- Centers for Disease Control and Prevention
CRS	- Children's Rehabilitative Services (ADHS)
ICD	- International Classification of Disease
MACDP	- Metropolitan Atlanta Congenital Defects Program

APPENDIX 5

Exclusion List - ABDMP
Non-reportable Birth Defects Cases

The following potential cases are not included in the ABDMP report for 1991:

- Duplicate abstracts and/or duplicated anomalies (cases with multiple abstracts; child seen at more than one facility), i.e., duplicate cases are merged and counted once.

- "Possibles" abstracted for review and consideration and subsequently determined to have conditions or defects that were not reportable - referring to CDC and CBDMP list of `excludables.

- Babies born to mothers whose residence is out-of-state or out-of-country (i.e., nonresident cases).

- "Negatives," that is of cases ruled-out during case finding and medical record review.

- "No Match" cases: Birth Certificate was not on file and state of birth cannot be confirmed as Arizona.

- Cases among aborted fetuses less than 20 weeks gestation and weighing less than 500 grams. These cases were excluded because there is no reliable denominator that can be used to generate a birth defect rate.

- Prenatally diagnosed cases that have not resulted in a live birth or stillbirth are not included. The ABDMP is not currently visiting prenatal diagnostic centers to identify cases.

- Defects with a "precision of diagnosis" code 1-7 are excluded. Only those defects diagnosed at the higher levels of precision (8 or above)are included. Refer to Appendix 3 for list of Precision of Diagnosis codes.

- Cases only diagnosed outside of the hospital setting are not included in the ABDMP.

APPENDIX 6

References

1 Hexter AC, Harris JA, Roeper P, Croen LA, Krueger P, Gant D (1990). Evaluation of the hospital discharge diagnoses index and the birth certificate as sources of information on birth defects. Public Health Reports 105:296-306.

2 Anderson RN, Kochanek KD, Murphy SL (1997). Report of final mortality statistics, 1995. Monthly Vital Statistics Report Vol. 45; Supplement 2; June 12.

3 Waitzman NJ, Romano PS, Scheffler RM, Harris JA (1995). Economic costs of birth defects and cerebral palsy - United States, 1992. Morbidity and Mortality Weekly Report Vol. 44; No. 37:695-699.

4 Keefer S, Rickard R (1994). Birth Defects and Developmental Disabilities in Colorado 1989-1992. Colorado Registry for Children with Special Needs.

5 Stierman L (1996). Birth Defects in Eleven California Counties: 1990-1992. California Birth Defects Monitoring Program.

6 Ibid.

7 Torfs CP, Katz EA, Bateson TF, Lam PK, and Curry CJ (1996). Maternal medications and environmental exposures as risk factors for gastroschisis. Teratology 54:84-92.

APPENDIX 7

Birth weight

The Arizona Birth Defects Monitoring Program monitors the distribution of birth weight. The data is obtainable from the birth certificate and may allow the detection of major shifts over time in the proportion of newborns with low birth weight.